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# Measurement of Physical Activity in Adults with Muscular Dystrophy: A Systematic Review

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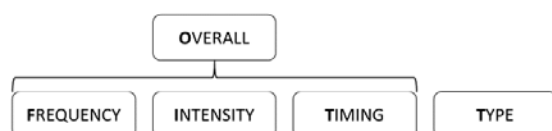
**Abstract** There is little consensus about measurement of physical activity in adults with muscular dystrophy. This systematic review summarizes evidence for measurement properties of direct and indirect measures of physical activity in adults with muscular dystrophy. A two-phase search for peer-reviewed articles identified firstly, studies which measured physical activity in this population and secondly, studies reporting the measurement properties of activity measures. Methodological quality was assessed using COSMIN guidelines and a best evidence synthesis conducted. Phase 1 included 53 studies identifying 63 measures including accelerometers, direct observation, heart rate monitors, calorimetry, positional sensors, activity diaries, single scales and questionnaires. Phase 2 included 26 studies of measurement properties for 32 measures. Methodological quality of the included studies was low, only 2 were rated good. There was insufficient evidence to robustly recommend any physical activity measures and further research is required to validate measures of physical activity for adults with muscular dystrophy. Based on the findings of this review, measures with potential for further study have been highlighted.

**Keywords:** *measure, physical activity, muscular dystrophy, adult, validity*

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## 1. Introduction

The aim of this review was to appraise measures of physical activity for the assessment of adults with muscular dystrophy. Effective physical activity measurement is important to evaluate outcomes in randomised controlled trials (RCTs), to monitor disease progression and to make recommendations for optimising physical activity [1]. For adults with muscular dystrophy, physical activity has been linked to health benefits, such as improved fitness and self-management [2,3]. However, more research reporting quantified physical activity levels is required to determine optimal activity for adults with muscular dystrophy and to evaluate potential risks, such as exercise-induced damage to dystrophic muscle [4,5].



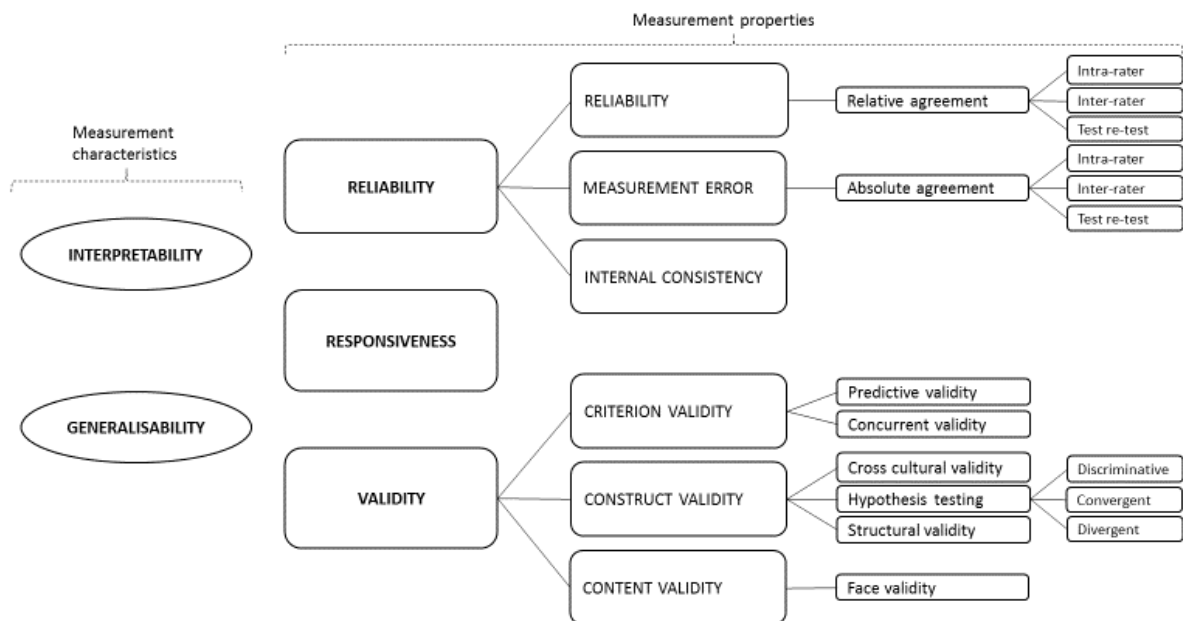
**Figure 1.** FITT framework for measurement of physical activity. Diagram created by author (SRL) based on common clinical practice and physical activity measurement analysis frameworks applied in the literature [8,9]

Physical activity is defined as behaviours involving bodily movements and energy expenditure [6,7]. Measurement of physical activity can be defined using a well-recognised

conceptual framework [8,9] which considers Frequency, Intensity, Timing and Type (FITT) of activity or overall measurement encompassing these parameters (see Figure 1).

The qualities of measurement tools can be defined in terms of measurement characteristics and properties, according to the Consensus-based Standards for the selection of health Measurement INstruments (COSMIN) taxonomy [10] (see Figure 2).

Measurement characteristics (generalizability and interpretability) of physical activity measures are variable because they depend on population and setting, and because there are numerous diverse ways to measure physical activity. These include indirect self-report tools, such as diaries and questionnaires, and direct tools which record the physiological consequences of activity, including bodily movements, metabolism and cardio-respiratory responses [11,12]. The characteristic pros and cons of these measurement tools (such as ease of use, burden, range and ability to capture FITT parameters) have been discussed in healthy individuals [8,12,13,14,15] older adults [9], wheelchair users [16] and people with neuromuscular diseases [17]. However, it is not known which measures might be most suitable for the assessment of physical activity specifically in adults with muscular dystrophy which is characterised by progressive weakness, heterogeneous presentations and variable function. It is therefore important to ascertain the generalizability and interpretability of physical activity measures in adults with muscular dystrophy to aid selection of appropriate measurement tools.



**Figure 2.** Framework of measurement characteristics and measurement properties. Diagram created by author (SRL) based on COnsensus-based Standards for the selection of health Measurement INstruments (COSMIN) checklist [28] and manual taxonomy and definitions [10]

The measurement properties (reliability, responsiveness and validity) of physical activity measures have also been investigated in multiple studies in various populations, including other neurological, rheumatological, oncological or pulmonary conditions and healthy, elderly, disabled or cognitively impaired individuals [9,12,13,17]. However, cumulative evidence is inconclusive due to conflicting reports, varied study design, diversity of measures and lack of consensus about gold standard criterion measures. Furthermore, reliability, responsiveness and validity of physical activity measures established in other populations may not be transferable to adults with muscular dystrophy who may have very different muscle, metabolic and cardiac functioning [18,19,20,21]. Thus, the measurement properties of physical activity measures when used with adults with muscular dystrophy remain unclear. To the authors' knowledge, this is the first review to examine population specific evidence for the reliability, responsiveness and validity of physical activity measurement in adults with muscular dystrophy.

The objectives of this review were, firstly, to identify direct and indirect physical activity measures used to assess adults with muscular dystrophy in a range of study designs and to describe their generalizability and interpretability. Secondly, to appraise the evidence of reliability, responsiveness and validity for physical activity measures in studies which included adults with muscular dystrophy. Finally, based on a narrative synthesis, to make recommendations, where possible, for the selection of suitable physical activity measurement tools for use with adults who have muscular dystrophy.

## 2. Methods

The protocol was registered on PROSPERO in July 2017 (Registration Number CRD42017070514) and follows the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines [22].

### 2.1. Search

The search was conducted in 2 phases. Phase 1 was designed to identify physical activity measures used to assess adults with muscular dystrophy and to describe their generalizability and interpretability. Phase 2 was designed to identify and appraise evidence for their reliability, responsiveness and validity.

In July 2017 (phase 1) and January 2018 (phase 2) the following databases were searched: Medline (1946-2017/8), Embase + Embase Classic (1947-2017/8), Global Health (1973-2017/8), PsycINFO (1806-2017/8), HMC Health Management Information Consortium (1979-2017/8) and Journals @ Ovid Full Text. In phase 1, Ovid search terms were expanded from 'muscular dystrophy', 'physical activity' and 'measure'. In phase 2, the search strategy was informed by the previous search results and additional 'measurement property' search terms were added. (For full search see supplementary material appendix I.)

#### 2.1.1. Study Selection

Studies were selected by 3 reviewers using the eligibility criteria listed in Table 1. (In phase 1: SRL and CW (10% sample); in phase 2: SRL, FS and CW (10% sample)). Disagreements were resolved by consensus discussion at this stage and throughout (arbitrated by CW).

Studies of any design were included if they had measured physical activity in any adult(s) with muscular dystrophy. Only studies where the measurement of activity spanned more than 10 minutes were included as shorter bouts of activity are not considered to contribute to recommended daily activity tallies [23,24,25]. The FITT framework (see Figure 1) was applied to ensure that only those intending to quantify physical activity overall or in 3 or more FITT parameters were included. When several reports pertained to the same study, the most recent or comprehensive article in terms of physical activity measurement was selected. In phase 2, inclusion was further limited to full text articles that evaluated reliability,

responsiveness and validity. A scarcity of physical activity measurement evaluation studies in adults with muscular dystrophy was anticipated. So, inclusion encompassed not only studies that overtly reported validity, reliability and responsiveness but also those that included hypothesis testing which incidentally indicated measurement properties of physical activity measures.

### 2.1.2. Data Extraction

The data extraction form was developed a priori, customised from previously published extraction tools [26,27] (see supplementary material appendix II). In phase 1, descriptive data were extracted by a single reviewer (SRL). In phase 2, 2 reviewers (SRL and FS) independently extracted the data.

### 2.1.3. Methodological Quality

In phase 1, methodological quality assessment was unnecessary because data were descriptive only. In phase 2, methodological quality was independently assessed by

2 reviewers (SRL and FS) using the COSMIN guidelines [28] to rate evidence supporting measure reliability, responsiveness or validity as excellent, good, fair or poor.

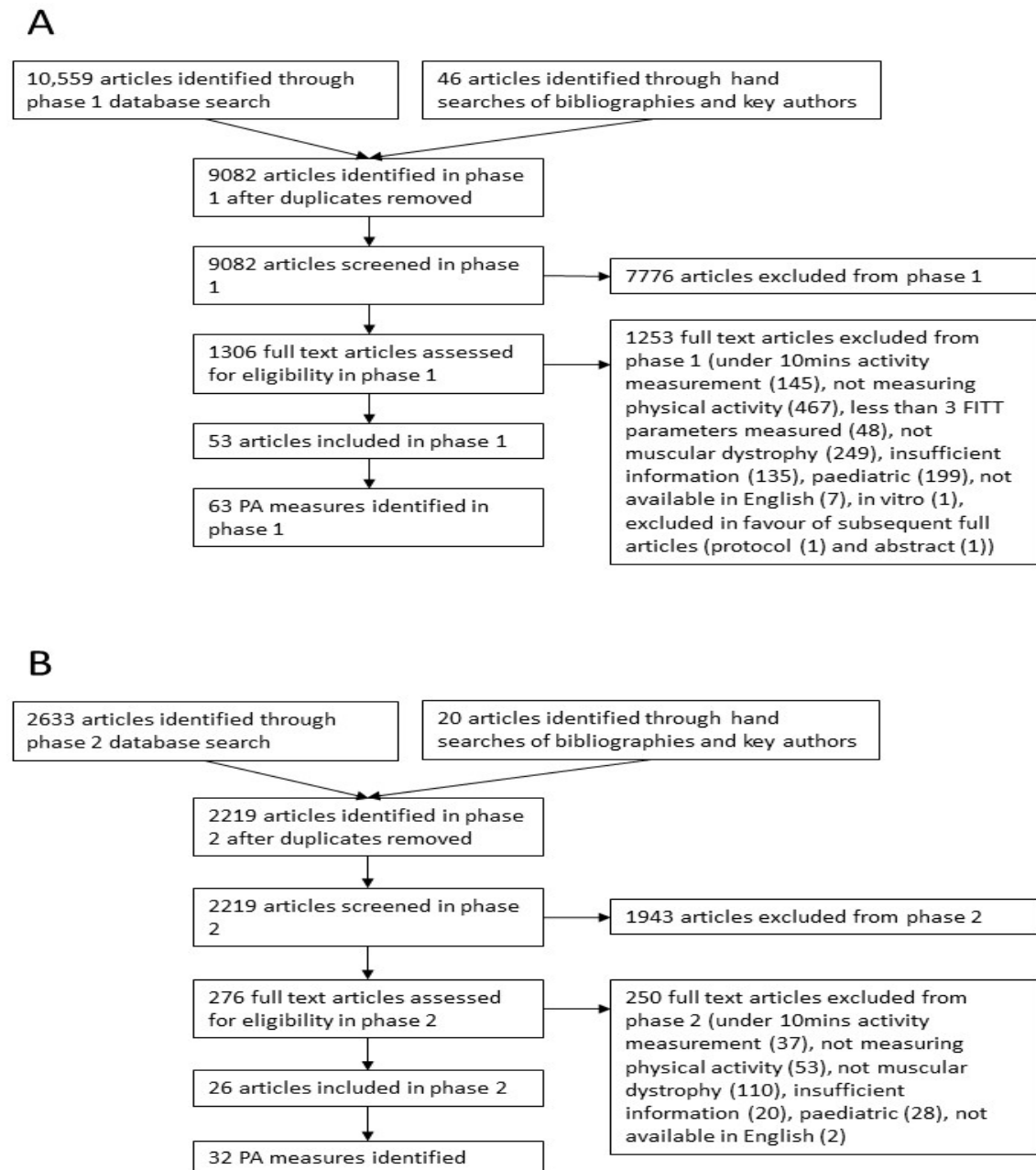
### 2.1.4. Synthesis

In phase 1, physical activity measures identified were described, listed and categorised. Their generalizability was quantified in terms of number of studies, number of participants, demographics (including age range, gender, diagnoses and mobility) and environment. Their interpretability was considered in terms of FITT measurement scope (i.e. capture of how often and how long different activities were carried out for and at what intensity, for example light, moderate or vigorous), timeframe, mode, metric and range of scores. In phase 2, evidence, and its methodological quality rating, for the validity, reliability and responsiveness of physical activity measures was listed for each included study. A narrative synthesis was carried out, considering the strength and consistency of evidence.

**Table 1. Inclusion and exclusion criteria**

Inclusion	Exclusion
<b>Participants</b>	
Study cohorts including at least 1 adult (aged over 18 years*) with a diagnosis of Muscular Dystrophy including: <ul style="list-style-type: none"> <li>• Limb Girdle Muscular Dystrophy (LGMD)</li> <li>• Myoshi Myopathy (MM)</li> <li>• Becker's Muscular Dystrophy (BMD)</li> <li>• Fascioscapulohumeral Muscular Dystrophy (FSHD)</li> <li>• Oculopharangeal Muscular Dystrophy (OPMD)</li> <li>• Myotonic Dystrophy or Dystrophia Myotonica (DM)</li> <li>• Unspecified Muscular Dystrophy (MD)</li> <li>• Duchenne Muscular Dystrophy (DMD)</li> <li>• Adult Congenital Muscular Dystrophy (CMD)</li> </ul>	Non-human subjects or in vitro cell cultures Cognitive impairment, pediatric* Other causes of muscle wasting including: <ul style="list-style-type: none"> <li>• Inflammatory myopathies</li> <li>• Metabolic myopathies</li> <li>• Other neuromuscular conditions (e.g. Spinal Muscular Atrophy, bulbospinal muscular atrophy (Kennedy's disease), Cerebral Palsy, Spina Bifida, Multiple Sclerosis)</li> <li>• Muscle wasting secondary to other conditions (e.g. Amputation, Cancer, Stroke, Spinal Cord Injury, Rheumatoid Conditions, immobility)</li> </ul>
<b>Intervention</b>	
Physical activity including free-living activities or prescribed exercise, lasting 10 minutes or more, where physical activity can be defined as: <ul style="list-style-type: none"> <li>• "Behaviour that involves human movement, resulting in physiological attributes including increased energy expenditure and improved physical fitness" (page S11) [7] or</li> <li>• "Any bodily movement produced by skeletal muscles that requires energy expenditure beyond resting expenditure" (page 3109) [6] and "the execution of a task or action by an individual" (page 577) [121]</li> </ul>	No physical activity The following types of activities: <ul style="list-style-type: none"> <li>• activity lasting less than 10 minutes (or for an undisclosed time period)</li> <li>• Activities during therapy sessions</li> <li>• Activities performed at relative rest (e.g. sleep or nocturnal movements, resting activity, mouth or tongue exercises, small muscle hand exercises and pulmonary muscle training)</li> <li>• Functional activity milestones (e.g. interval analysis of loss of ambulation)</li> </ul>
<b>Comparison and outcomes</b>	
Studies reporting measurement of physical activity. Measurement encompassing 3 or more FITT parameters by a battery of measurement approaches or encompassing overall physical activity measurement for: <ul style="list-style-type: none"> <li>• An evaluation of measurement properties</li> <li>• A primary or secondary outcome</li> <li>• Assessment of compliance</li> <li>• An independent variable</li> <li>• A demographic characteristic</li> </ul>	Studies with no measurement of physical activity reported, or measurement period lasting under 10mins. Studies reporting only about measures: <ul style="list-style-type: none"> <li>• Of activity limitation i.e. ease/difficulty of performing activities (including quality of life measures)</li> <li>• Of fitness (e.g. from a single time point, such as maximal oxidative capacity, maximal workload or exertion rating)</li> <li>• Used to capture only resting activity without corresponding active measurement</li> </ul>
<b>Study Design</b>	
Peer reviewed articles† of any methodological design including: <ul style="list-style-type: none"> <li>Measure evaluation studies</li> <li>Experimental studies (randomised designs)</li> <li>Observational analytic studies (pre/post, comparison)</li> <li>Descriptive (longitudinal, cross-sectional, case studies)</li> <li>Prospective or retrospective</li> </ul>	Bibliographies of relevant full texts were hand searched for pertinent articles and authors, but the following were excluded: <ul style="list-style-type: none"> <li>Reviews (systematic or narrative)</li> <li>Journal letters/ correspondence/ consensus summaries</li> <li>Unpublished data</li> <li>Articles not available in English language</li> </ul>

\* If age range was not reported, a conservative estimated range was extracted as mean  $\pm$  1 standard deviation to account for skewed distribution towards lower ages in pediatric studies. † In phase 1 abstracts and protocols were included unless superseded by a full text article; in phase 2 only full text articles were included.



**Figure 3.** Study selection schematics for phase 1 (A) and phase 2 (B)

## 3. Results

### 3.1. Phase 1

Study selection is summarised in Figure 3a. Agreement between reviewers (SRL and CW) was 90% and 87% for abstract and full text screening respectively, with full agreement after consensus discussion. Included articles are listed and described in Table 2; 63 physical activity measures were identified (see Table 3 and 3a supplement).

#### 3.1.1. Generalizability

Activity measurement was generalizable across gender, mobility from independent walking to wheelchair use, age range from teenager to elderly and amongst different muscular dystrophy diagnoses. Myotonic Dystrophy and Facioscapulohumeral Dystrophy were the most commonly assessed. Indirect measures were used to assess larger

numbers of participants than direct measures, particularly standardised questionnaires ( $n=1567$ ). Of the direct measures, the greatest number of participants were assessed using accelerometry ( $n=731$ ). Many studies used standardised questionnaires, non-standardised diaries, heartrate monitoring, direct observation and accelerometry (15 studies [29-43] including 3 RCTs [29,33,41], 18 studies [29,33,40-41,44-57] including 4 RCTs [33,41,44,48], 15 studies [29,33,40,44-45,48-49,50-55,58-59] including 5 RCTs [29,33,44,48,58], 13 studies [29,44,48,51,58-66] including 4 RCTs [29,48,58,65] and 13 studies [29,33,37,41,44,46,53,61,67-71] including 6 RCTs [29,33,41,44,67,68] respectively). Whereas, single and sub-scale activity measures, calorimetry and positioning systems were the least widely used in the fewest participants.

Free-living physical activity was most usually assessed, especially by questionnaires, diaries, continuous heartrate monitoring and accelerometry. However, prescribed



activities were also assessed at home and in other environments, including the gym, sports pitch and laboratory, where activity was monitored by indirect calorimetry, periodic heartrate monitoring and some training logs.

The most generalizable tools within each category, used in the most studies and participants with the

widest spectrum of demographics, included 2 standardised questionnaires (the International Physical Activity Questionnaire (IPAQ), the Physical Activity Scale for Individuals with Physical Disabilities (PASIPD)), activity logs, Polar heartrate monitors and triaxial, ankle accelerometers (although, the only accelerometer used to assess non-ambulant participants was wrist-worn [46]).

**Table 2. Description of 53 included studies**

Design (n) / Description*	Citation	Condition (n)	Age range	Gender (m/f)	Phase
<b>Experimental (9)</b>					
RCT	Andersen et al., 2017 [29]	FSHD (12)	26-67	9/3	1 & 2
	Andersen et al., 2015a [33]	FDHD (41)	22-65	21/20	1
RCT (crossover)	Voet et al., 2014 [44]	FSHD (77)	20-79	37/40	1 & 2
	van der Kooi et al., 2007 [65]	FSHD (65)	27-51†	40/25	1 & 2
RCT (protocol)	Veenhuizen et al., 2015 [58]	FSHD (estimated 17)	N/R	N/R	1
	Van Engelen et al., 2015 [68]	DM (296)	N/R	N/R	1
RCT (pilot)	Kierkegaard et al., 2011b [48]	DM (35)	20-65	15/20	1 & 2
	Dawes et al., 2006 [41]	BMD (4), FSHD (3), LGMD (4), CMD (1), DM (4)	18-81	10/6 ‡	1 & 2
RCT (2° analysis)	Janssen et al., 2016 [67]	FSHD (31)	39-67†	22/9	1 & 2
<b>Observational analytic (26)</b>					
Cross-sectional	Jacques et al., 2017 [30]	BMD (21)	26-52†	21/0	1 & 2
	Heutink et al., 2017 [72]	DMD (estimated 30)	16-26†	30/0 ‡	1 & 2
	McCorry et al., 1998 [55]	DM (9), FSHD (3), BMD (1)	29-63†	8/5 ‡	1 & 2
Cohort	Brun et al., 2017 [122]	LGMD (41)	12-72	20/21	1
	Smith et al., 2016 [31]	MD (282)	N/R	N/R	1 & 2
	Morse et al., 2016 [32]	FSHD (11), BMD (11), LGMD (9), DMD (11)	18-59	42/0	1 & 2
	Andersen et al., 2015b [62]	DM (7), FSHD (6), LGMD (2), BMD (2)	18-52	10/7	1
	Vissing et al., 2014 [45]	LGMD (4), MM (2)	39-54	4/2	1
	Dahlqvist et al., 2014 [63]	BMD (4)	26-32	4/0	1
	Andersen et al., 2013 [64]	BMD (5), FSHD (5), LGMD (4)	20-58	12/2	1 & 2
	Sveen et al., 2008 [49]	BMD (11)	28-36†	11/0	1
	Sveen et al., 2007 [50]	LGMD (9)	43-53†	6/3	1
	Wintzen et al., 2007 [123]	DM (13)	29-57†	5/8	1
	Wiles et al., 2006 [70]	DM (13)	44-48†	N/R	1 & 2
	Aitkens et al., 2005 [54]	DM (4), LGMD (3), FSHD (1)	47-54†	N/R	1 & 2
	Kilmer et al., 2005 [53]	LGMD (8), DM (4), FSHD (2), BMD (2)	37-63†	10/6 ‡	1 & 2
	Orngreen et al., 2005 [52]	DM (12)	21-58	9/3	1
	Olsen et al., 2005 [51]	FSHD (8)	18-55	4/4	1
	Taivassalo et al., 1999 [66]	LGMD (4), MD (3), OPMD (2), DM (1)	29-63	5/5	1
	Wright et al., 1996 [56]	DM (5), LGMD (1)	28-40	4/2	1
	Aitkens et al., 1993 [57]	DM (12), LGMD (3), FSHD (2)	25-55†	11/6 ‡	1
	Florence and Hagberg, 1984 [74]	LGMD (3), CMD (2), FSHD (1)	21-40	6/0	1
Cohort (protocol)	Lassche et al., 2013 [37]	FSHD (predicted 14), OPMD (predicted 14)	N/R	N/R	1
Case control	Brady et al., 2014 [124]	DM (63)	26-57†	26/37	1
	Angelini et al., 2011 [125]	MM (13), LGMD (3)	16-50	12/4 ‡	1
	Kalkman et al., 2007 [69]	DM (70), FSHD (60)	22-61	75/55 ‡	1 & 2
<b>Descriptive (18)</b>					
Cross-sectional (Evaluative)	Martin and Whalen, 2012 [75]	MD (4)	13-64†	2/2 ‡	1 & 2
	Busse et al., 2004 [71]	MD (10)	39-65†	6/4	1 & 2
	Washburn et al., 2002 [42]	MD (estimated 25)	N/R	N/R	1 & 2
	Kimura et al., 2014 [46]	DMD (12)	10-19	12/0	1 & 2
	Chen et al., 2014 [47]	MD (1)	46	0/1	1 & 2
Cross-sectional	Barfield et al., 2016 [60]	MD (4)	7-63	N/R (4)	1 & 2
	Matsuda et al., 2015 [34]	MD (332)	18-65	140/192	1
	Rosenberg et al., 2013 [36]	MD (321)	20-85	136/185	1 & 2
	Shimizu-Fujiwara et al., 2012 [126]	DMD (6)	18-24	6/0	1
	Komaki et al., 2011 [127]	DMD (26)	14-38	26/0	1
	Kierkegaard et al., 2011a [38]	DM (70)	19-70	29/41	1 & 2
	Harris et al., 2010 [73]	MD (1)	36-50	N/R	1
	Phillips et al., 2009 [40]	FSHD (5), DM (5), LGMD (1)	32-56†	3/8 ‡	1 & 2
	Longmuir and Bar-Or, 2000 [43]	MD (12)	15-20	6/6	1 & 2
Cross-sectional abstracts	Eichinger et al., 2013 [35]	DM (107)	N/R	57/50	1
	Phillips and Marr, 2009 [39]	DM (97), LGMD (82), FSHD (68)	24-65†	N/R	1
	Meilleur et al., 2015 [61]	CMD (11)	N/R	N/R	1
Case study	Pasotti et al., 2014 [59]	FSHD (1)	43	0/1	1

\*Classified according to Centre for Evidence-Based Medicine study design guidelines [128]. † Range calculated by mean (or median)  $\pm$  1 standard deviation (or confidence interval) in the absence of reported range. ‡ Estimated age range from proportion of larger sample. Abbreviations: RCT – Randomized Controlled Trial; FSHD – Facioscapulohumeral Dystrophy; DM – Dystrophia Myotonica (Myotonic Dystrophy); BMD – Becker's Muscular Dystrophy; DMD – Duchenne's Muscular Dystrophy; LGMD – Limb Girdle Muscular dystrophy; MD – Muscular Dystrophy (type unspecified); MM – Miyoshi-like Myopathy; OPMD – Oculopharyngeal Muscular Dystrophy; CMD – Congenital Muscular Dystrophy; N/R – not reported.

**Table 3. List of 63 measures used to assess adults with muscular dystrophy**

<b>Indirect Measures (27)</b>	
IPAQ	International Physical Activity Questionnaire [29,31,34-37]
PASIPD	Physical Activity Scale for Individuals with Physical Disabilities [32,39,42]
B-PAR	Bouchard Physical Activity Record [33]
EPIC	Norfolk Physical Activity Questionnaire [40]
PASE	Physical Activity Scale for the Elderly [41]
GLTEQ	Godin Leisure Time Exercise Questionnaire [36]
CFS	The Canada Fitness Survey [43]
BPAQ	Bone-specific Physical Activity Questionnaire [30,32]
FAI	Frenchay Activities Index [38]
PSDQ – activity	Subscale of the Physical Self-Description Questionnaire [75]
ACS – participation	Subscale/specific ranking of Activity Card Sort [58]
FPACQ – transport	Subscale of the Flemish Physical Activity Computerized Questionnaire [72]
MAQ – activity	Subscale of the Modifiable Activity Questionnaire [72]
PAGS	Physical Activity Grading Scale (single scale) [38,48]
ACR	Activity Change Rating (single scale) [51-52]
AI 1-5	Activity Increment 1-5 (single scale) [123]
DOA	Daily Observation of Activity (single scale) [65]
Diary	Physical activity recorded in a diary [40,46,54,55]
Diary (electronic)	Physical activity recorded in computer software [47]
Dairy + DQ	Physical activity recorded in a diary with a direct quantifier (step count) [53]
Log	Training log [29,33,48-49,51-52]
Log + Q	Training log with a self-report quantifier (rating of perceived exertion) [41]
Log + D	Training log with direct quantifiers (heartrate, revolutions per minute) [45,50]
Log + Q + D	Training log with rating of perceived exertion and direct quantifiers (HR, workload) [44,56]
Log + Q ('phone)	Training log by telephone with self-report quantifiers (rating of perceived exertion and repetitions) [57]
Interview	Study specific interviews [33,73,124,125]
Survey	Study specific survey [122]
<b>Direct Measures (36)</b>	
Pedometer	Unspecified (waist) [41]
Pedometer	Yamax Digiwalker (waist, uniaxial) [53]
Pedometer	Omron Walking Style Pro (waist, triaxial) [29]
Actometer	Unspecified (ankle) [37]
Actometer	StepWatch™ (ankle, biaxial) [70-71]
Actometer	Actilog V3.0 (ankle, triaxial) [44,67,69]
Actometer	Motionlogger Watch (wrist, triaxial) [46]
Actometer	Actigraph GTX3 (wrist, triaxial) [61]
Actometer	Kinesense GeneActiv (wrist, triaxial) [68]
Actometer	SenseWear Pro (arm, biaxial plus temperature sensing) [33]
Pulsewatch	Unspecified [33,58]
Pulsewatch	Polar (model not specified) [51-52]
Pulsewatch	Polar RS400 [59]
Pulsewatch	Polar (S610) [29,48]
Pulsewatch	Polar (Accurex) [49-50]
Pulsewatch	Polar (FT40) [45]
Pulsewatch	Polar (Vantage XL) [53-55]
Pulsewatch	Garmin Forerunner 50 [44]
Heart monitor	Polar (model not specified) [40]
Counting	Directly observed step counts [61]
Spectating	Directly observed competitive sport [60]
Video + D	Supervision via Skype with a direct quantifier (workload) [59]
Supervision	Directly observed supervised exercise [29,51,58,65]
Supervision + R	Directly observed exercise with self-report quantifier (rating of perceived exertion) [48]
Supervision + D	Directly observed exercise with a direct quantifier (heartrate) [66,74]
Supervision + R + D	Directly observed exercise with rating of perceived exertion and direct quantifiers (heartrate, workload) [44,62,63,64]
Indirect calorimetry	Gaseous exchange analyser (K4b2) [60]
Indirect calorimetry	Gaseous exchange analyser (Oxycon Mobile) [60]
Indirect calorimetry	Gaseous exchange analyser (Quark b2, Cosmed) [64]
Indirect calorimetry	Gaseous exchange analyser (2900 Metabolic System) [53-55]
TEE by DLW	Total Energy Expenditure calculated using Double -Labelled Water [127]
TEE by diet	Total Energy Expenditure calculated using dietary intake [126-127]
TEE by HR	Total Energy Expenditure from monitoring heartrate, oxidative capacity equivalence and factorial diaries [53-55]
PAL	Physical Activity Level index calculated from total / resting energy expenditure [53,54,126,127]
GPS	UbiTrak Global Positioning System tracker [47]
GPS	Participation and Activity Measurement Global Positioning System tracker [73]

For full measure descriptions and categorizations, see Table 3a in the supplementary material.

### 3.1.2. Interpretability

Indirect measures collected activity spanning 3 days to a year (or lifetime), some in real-time, including activity diaries of 3 days to 6 months, and others by recall, including standardised questionnaires often over 7 days. Whereas, all direct measures recorded activity in real-time from 10 minutes to 6 months. Most recording periods lasted 1-14 days, except for direct observation, periodic heart rate monitoring and indirect calorimetry which were conducted over shorter timeframes of between 10-90 minutes. There was great variability in the metrics of activity measures, making it difficult to compare activity measurement ranges (see table 3 and 3a supplement).

Standardised questionnaires and diaries had the best scope to quantify discrete FITT parameters; whereas direct measures like accelerometry and energy expenditure calculations were usually concerned with overall physical activity.

Interpretability was boosted by using multiple physical activity measures in 32 included studies [29,32-33,36-38, 40-41,44-46,48-60,62-66,72-74], including 7 of the 9 RCTs [29,33,41,44,48,58,65]. Multiple measures increased the scope of physical activity measurement in terms of FITT in 29 studies [29,33,38,40-41,44-46,48-60,62-66,72-74] and ocomplementary indirect and direct measures were employed in 29 studies [29,33,37,40-41,44-58, 60, 62-66,72-74].

**Table 4. Study characteristics of studies and evaluated measures included in phase 2**

Author / COSMIN	Study Description	Measure(s)	Measurement properties
Washburn et al., 2002  GOOD	Observational descriptive cross-sectional study for measure evaluation of the Physical Activity Scale for Individuals with Physical Disabilities in 372 disabled people [42]	PASIPD  (Physical Activity Scale for Individuals with Physical Disabilities)	<u>Reliability</u> Internal consistency was demonstrated by significant item correlations with total score ( $r=0.20-0.67$ for all items, $p<0.05$ ) and factor analysis of 5 clusters (accounting for 63% of variance in total scores, with Cronbach's alpha ranging from .37 to .65) <u>Construct validity</u> Discriminative validity was demonstrated by extreme group differences in PASIPD score between young versus old (mean difference = 8.1, $p<0.001$ ), self-rated health excellent versus fair/poor (mean difference = 11.9, $p<0.001$ ), attendant care versus those without (mean difference = 11.9, $p<0.001$ ) and self-rated physical activity (mean difference = 17.5, $p<0.05$ )
Martin and Whalen, 2012  FAIR	Observational descriptive cross-sectional study for measure evaluation of the Physical Self-Description Questionnaire in 50 disabled people [75]	PSDQ – Activity  (Physical activity subscale of the Physical Self-Description Questionnaire)	<u>Reliability</u> Internal consistency of subscale was high (Cronbach's alpha =0.85) <u>Construct validity</u> Convergent validity demonstrated by significant moderate to low associations with certain PSDQ subscales (sport confidence $r=0.56$ , flexibility $r=0.50$ , coordination $r=0.43$ , endurance $r=0.33$ (all $p<0.05$ ), strength $r=0.62$ and multiple regression analysis $\beta=0.37$ , $r^2=0.47$ , $p=0.03$ ). Convergent validity was not demonstrated with the other subscales of the PSDQ.
Kimura et al., 2014  POOR	Observational descriptive cross-sectional study for measure evaluation of the utility of a wrist actigraph for estimating muscle strength in 22 people with Duchenne's Muscular Dystrophy [46]	Actometer  (Motionlogger Watch; triaxial wrist actigraph, Ambulatory Monitoring, Ardsley, NY, USA)	<u>Construct validity</u> Discriminative validity was demonstrated by extreme group differences in Actigraph zero crossing mode (ZCM) and Actigraph proportional integration mode (PIM) scores between ambulant and non-ambulant people with DMD (Wilcoxon rank-sum test, ZCM $p<0.001$ , mean difference 44 counts per minute, PIM $p<0.001$ , mean difference 36406 bits) Convergent validity was demonstrated by weak to moderate significant correlations of Actigraph ZCM and PIM scores with 6-minute walk distance (6MWD) and knee extension strength measured using handheld dynamometry (Spearman's, ZCM with 6MWD, $\rho = -0.44$ , $p<0.01$ , ZCM with knee extension strength, $\rho = 0.25$ , $p<0.01$ , PIM with 6MWD, $\rho = 0.58$ , $p<0.01$ , PIM with knee extension strength, $\rho = 0.63$ , $p<0.01$ )
Chen et al., 2014  POOR	Observational descriptive cross-sectional study for measure evaluation of a novel movement tracking system in 5 disabled people [47]	UbiTrak (GPS)  (Global Positioning System and Wireless Fidelity tracker)	<u>Reliability</u> Measurement error was demonstrated as acceptable by good to strong agreement between UbiTrak and electronic activity diaries (62-87% agreement)
Busse et al., 2004  POOR	Observational descriptive cross-sectional study for measure evaluation of the StepWatch in 10 people with primary muscle disease and comparison to healthy controls [71]	Actometer  (StepWatch™, biaxial, ankle step activity monitor (Cymatech, USA))	<u>Reliability</u> Test-re-test reliability demonstrated for 7-day mean step count 1-3 weeks apart (ICC=0.86) and for peak 30-minute step count and average 20, 30 and 60-minute step counts (ICC=0.82-0.95) <u>Construct validity</u> Discriminative validity was demonstrated by extreme group differences in mean 7-day step count between mobility impaired and healthy (independent t-test, mean difference = 2365, $p=0.001$ ) Convergent validity was demonstrated by significant correlation between gait speed and mean 7-day step count (Pearson's, $r = 0.45$ , $p=0.01$ ) but not demonstrated with the Rivermead Mobility Index.

For incidental measurement properties see table 4a in the supplementary material. Abbreviations: ICC – intraclass correlation coefficient; DMD – Duchenne's Muscular Dystrophy; ZCM – zero crossing mode; PIM – proportional integration mode; 6MWD – 6-minute walk distance.



### 3.2. Phase 2

Study selection is summarised in Figure 3b. Agreement between the 2 reviewers (SRL and FS) was 86%, 87%, 91% and 86% for abstract, full text screening, data extraction and COSMIN ratings respectively, with full agreement after consensus discussion. Evidence for the reliability, responsiveness and validity of 32 physical activity measures is listed in Table 4 (and 4a supplement). Only 5 included studies [42,46,47,71,75] had as their primary objective the evaluation of measurement properties of a physical activity measure; the remaining 21 articles were included for incidental measurement properties from hypothesis testing relating to other objectives. No studies were rated as excellent; 2 were rated as good [36,42], 12 as fair and 11 as poor. This was largely due to low sample sizes and incidental measure evaluation.

#### 3.2.1. Reliability and Responsiveness

There was very little evidence for reliability or responsiveness testing of any physical activity measures. Of the indirect measures, there was good quality evidence of internal consistency of the PASIPD from an evaluative study including 372 participants, an estimated 7% of whom had muscular dystrophy [42]. There was fair quality evidence of internal consistency of the Physical Self-Description Questionnaire (PSDQ-S) from an evaluative study including 50 participants, 8% of whom had muscular dystrophy [75]. There was also incidental report of moderate to high test-re-test reliability of the Canada Fitness Survey (CFS) [43,76].

Of the direct measures, there was poor quality evidence of good test-re-test reliability of the StepWatch [71] accelerometer and moderate measurement error of Ubitrak (a Wi-Fi and GPS (Global Positioning System) movement tracker) [47]. There was poor quality, incidental evidence of inter-rater reliability between indirect calorimetry gaseous analysers, K4 b2c and Oxycon Mobiled [60] and responsiveness of a pedometer compared to the Physical Activity Scale for the Elderly was tenuously indicated, as neither detected significant changes in physical activity post intervention [41].

#### 3.2.2. Validity

There was a small amount of evidence supporting the validity of 2 indirect measures (see table 4). The strongest evidence was for the PASIPD, which had good quality evidence of significant discriminative validity between extreme groups [42] and some incidental, consistent, fair quality evidence for concurrent validity [30,32]. There was also fair quality evidence for convergent validity of the PSDQ-S activity subscale [75]. Based on incidental findings only, there was some cumulative evidence for 2 other questionnaires: The Bone-specific Physical Activity Questionnaire (BPAQ), which had consistent, fair quality evidence for discriminative validity [30,32] and convergent validity [32] and the IPAQ, which had good quality evidence for convergent validity [36] and mixed, poor quality evidence for [29] and against discriminative validity [31]. Evidence for the validity of other direct measures was even more sparse. However, incidental validity of diaries was tentatively indicated, including low quality evidence of discriminative validity for an activity

diary [54,55] and convergent validity of a training log [48].

There was no good quality evidence supporting the validity of any direct measures. However, there was some collective, low-quality evidence concerning accelerometry and heartrate monitoring. There was cumulative, predominantly incidental, evidence of discriminative and convergent validity of accelerometry, which was stronger for triaxial accelerometers (Omron [29], Motionlogger [46] and Actilog [67,69]) than biaxial (StepWatch [70,71]) or uniaxial devices (Yamax digi-walker [53]) and for ankle [44,67,69,70,71] rather than trunk [29] or wrist [46] placement. There was incidental, consistent evidence for discriminative validity [40,54] and convergent validity [48] of Polar heartrate monitors and mixed evidence for [54,55] and against [53] discriminative validity of heartrate monitoring used with indirect calorimetry equivalence to estimate total energy expenditure.

## 4. Discussion

The main finding of this systematic review is that physical activity has been measured in numerous and various ways in a range of 53 studies assessing adults with muscular dystrophy. There is no consensus about the most generalizable or interpretable activity measurement tools for this group. Furthermore, evidence is limited about measure reliability, responsiveness and validity for the assessment of physical activity in adults with muscular dystrophy. Only 5 studies overtly evaluated the measurement properties of physical activity measures and none have provided high quality evidence of reliability, responsiveness and validity.

### 4.1. Direct Measures

Despite the paucity of evidence for reliability, responsiveness and validity of direct measures of physical activity in adults with muscular dystrophy, tools like accelerometry and heartrate monitoring might have potential. As demonstrated in the literature [9,14,15,16,77] and by the studies identified in this review, accelerometry and heartrate monitoring are both fairly generalizable and interpretable. Accelerometry can capture free-living activity over the medium (days/weeks) to long-term (months) and can detect frequency, absolute intensity, and timing, also yielding an overall quantification of physical activity. Although accelerometry cannot discern relative exertion or type of activity, it is adaptable, relatively inexpensive, and unobtrusive. In terms of measurement properties, tentative construct validity of accelerometry has been indicated in this review, with the best evidence in support of triaxial devices. Multi-plane movement detection, although not integral for regular walking, may be more suited to irregular torsions [78], characteristic of abnormal mobility in adults with muscular dystrophy [79]. Furthermore, for healthy people and those with chronic diseases, multi-axial devices have also demonstrated stronger criterion validity and lower measurement error than uniaxial devices [80]. Similarly, the triaxial GENEActiv has been validated over 6 minutes or less in adults with myotonic dystrophy [81] with construct validity supported incidentally in a high quality RCT [82] (too recent for inclusion in this systematic search) and the biaxial StepWatch has been

extensively validated in ambulant people with Multiple Sclerosis, Parkinson's Disease and children with Duchenne's Muscular Dystrophy [71,83,84,85]. In contrast, criterion validity was reportedly low and measurement error unacceptably high for the uniaxial Digi-walker over 2 minutes, in ambulant adults with neuromuscular diseases, including muscular dystrophy [86]. In this review, there was more evidence for generalizability of accelerometer placement on the ankle than the trunk or wrist, although it came only from ambulant participants; whereas, wrist placement better encompassed a range of mobility including wheelchair users [46]. In the literature, wrist accelerometry has been linked to non-ambulant assessment [87] and lower measurement error at slow walking speeds [88] which may become relevant as muscular dystrophy progresses [79]. Thus, triaxial accelerometry, placed at the ankle or wrist, represents a potential tool for the assessment of physical activity in adults with muscular dystrophy, subject to establishing robust reliability, responsiveness and validity in both ambulant and non-ambulant.

Heartrate monitoring may also have potential, particularly for monitoring compliance with, and recording intensity of, prescribed exercise interventions in adults with muscular dystrophy. In this review there were tentative indications for construct validity of Polar devices. They are generalizable and can record frequency, timing and relative intensity of exertion, which is particularly useful for quantifying prescribed activity [89]. However, heartrate monitors cannot differentiate between activity and emotional heartrate responses, thus there is a requirement to collect supplementary information, such as a diary or predetermined personal activity zone heartrates [54,55]. In addition, reduced heartrate variability in muscular dystrophies [20] might impact the interpretation of heartrate comparisons, especially relative to predicted values. Higher resting and lower submaximal/maximal heartrates have been reported in Duchenne's Muscular Dystrophy [83] and increased sympathetic drive with progressive parasympathetic dampening in Facioscapulohumeral Dystrophy [21]. Similar caution is required for energy expenditure extrapolations from heartrate or accelerometry data due to potentially altered metabolic functioning in adults with muscular dystrophy [18,19,90,91]. Thus, it is advisable to report actual recorded heartrate in beats per minute, or absolute step counts, and to treat extrapolated values with circumspection.

## 4.2. Indirect Measures

The same reservations about energy expenditure extrapolations must be applied to indirect measures that estimate metabolic expenditure. Additional caution is also necessary when interpreting questionnaire scores due to the potential for self-report, re-call and/or social desirability bias, which usually produce overestimations [8]. However, indirect, self-report measures of physical activity for adults with muscular dystrophy are widely generalizable, inexpensive, acceptable and easy to use [9,11,12].

Several standardized questionnaires were identified as having potential in this review. The PASIPD had the strongest evidence supporting its reliability and validity which is consistent with evidence from other populations including strong test-re-test reliability [92,93], discriminative validity [94] and low [92,93,95,96] to moderate [94]

criterion validity. However, significant overestimation measurement error has been reported [95]. In terms of interpretability, PASIPD comprehensively covers FITT and is sensitive to disabled and low-level activities, although it is unsuitable for comparisons with non-disabled populations. The IPAQ, BPAQ and PSDQ-S are suitable for comparison with other populations; the BPAQ and PSDQ-S are situation specific to bone health [97] and self-perception [98] respectively. The IPAQ is the most generalizable questionnaire identified in this review and various versions are available including short, long and modified versions (more sensitive to lower activity intensities and non-ambulant mobility [99,100]). In this review, the validity of the IPAQ was inconclusive. However, measurement properties established in other populations include strong test-re-test reliability [23,101,102] moderate responsiveness [103,104], low [23,100,101,103,104] to moderate [23,105] criterion and convergent [105] validity and predominantly overestimation measurement error [104,106]. Thus, if acceptable reliability, responsiveness and validity can be established and energy expenditure scores are treated circumspectly, both the PASIPD and IPAQ have potential for the assessment of physical activity in adults with muscular dystrophy.

Activity diaries also have potential as generalizable and interpretable activity measures, especially those designed to span FITT which are often used for prescribed activity monitoring. In addition, diaries might have potential as an adjunctive activity measure. Supplementary activity logs have been shown, for example, to mitigate IPAQ overestimation [107] and to improve criterion validity and measurement error [104]. Diaries have also been advocated alongside direct activity measures [14,15] and, in this review, diaries appeared to strengthen interpretation of heartrate monitoring and indirect calorimetry equivalence [53,54,55]. Activity diaries are, therefore, not only useful for monitoring prescribed activity, they may have an application as adjuncts to enhance interpretability of free-living physical activity measurement.

## 4.3. Implications

Clearly, all physical activity measures have limitations, both general and specific to adults with muscular dystrophy. These must be considered in study design and some authors have compiled checklists to aid measure selection [17,108]. There is also an argument, reflected by the findings of this review, for a multi-measurement approach, where multiple, complementary activity measures are employed, to improve the interpretation of physical activity measurement [14,15,16,80] and potentially improve measurement properties [104,107]. Recall bias can be neutralised by triangulation with real-time measurement and social desirability responding can be minimised by the knowledge that responses will be verified directly [109]. Recording both relative and absolute activity, by heartrate monitoring and accelerometry or GPS, can enrich physical activity data interpretation and has also been shown to improve measurement properties [110-113]. Thus, diaries, heartrate monitoring and, possibly, GPS might be suitable adjuncts to standardised questionnaires or accelerometry. A multi-measurement approach is recommended for the assessment of physical activity in adults with muscular dystrophy.

The current lack of research evaluating measurement properties of physical activity measurement in adults with muscular dystrophy means that authors should be encouraged to report study level reliability and validity of the measures employed in trials or observational studies.

In addition, measure evaluation studies are required to determine the validity, reliability and responsiveness of physical activity measures for use with adults with muscular dystrophy. The evidence, both evaluative and incidental, compiled in this review was predominantly low quality-rated, often linked to sample sizes below the 50-100 participant threshold set by COSMIN for high quality-ratings [27]. Sample size challenges include the rarity of adults with muscular dystrophy and restricting study designs to single diagnoses and/or separating ambulant and non-ambulant [1,17]. In larger samples, it is also difficult to find an activity measure suitable to encompass activity heterogeneity within and between muscular dystrophic diagnoses [114,115] and stages of disease progression [116,117,118]. Restrictive sampling is advocated for experimental designs [1]. However, to optimise statistical power, a larger, heterogeneous sample (with whole and sub-group analyses) is recommended for future evaluative studies where measurement properties are to be elucidated.

For evaluative research, it is also difficult to identify a gold-standard criterion measure of physical activity. In the wider physical activity literature, criterion measures include calorimetry, accelerometry and direct observation [8,12,13,119]. Due to burden and cost, direct observation and indirect calorimetry are limited to smaller samples and short timeframes (<1 day). Calorimetry by double-labelled water is suitable over a timeframe of 1-2 weeks, but burdensome. Energy expenditure calculations should also be viewed with caution because calorimetry is likely to be impacted by metabolic abnormalities and progressive physiological changes in muscular dystrophy [18,19,90]. Whereas, direct observation has inherent content validity [119] and, in this review, it was interpretable and generalizable in 13 studies. Thus, it represents a suitable, initial gold-standard criterion for short-term validation. Accelerometry is generalizable in larger samples and over various timeframes. Thus, accelerometry, with prior validation against direct observation, might represent a suitable criterion against which to validate other activity measures for adults with muscular dystrophy.

#### 4.4. Strengths and Limitations

To the authors' knowledge, this is the first systematic review about measurement characteristics and properties of physical activity measures specifically for adults with muscular dystrophy. The review employed a broad, sensitive search strategy, 3 independent reviewers and rigorous COSMIN appraisal. However, there are some limitations. These include, firstly, exclusion of non-English language articles which means relevant articles published in other languages may have been missed. Nevertheless, a recent review of physical activity measures in adults and children with neuromuscular diseases [17] did not identify additional measurement approaches beyond those identified in this review; which suggests no pertinent measures were missed. Secondly, there is potential for

bias in phase 1 as only a 10% sample was second reviewed and there was no methodological appraisal. However, the descriptive nature of phase 1 was straightforward, and the methodological quality of the studies did not impact description of the tool. Thirdly, COSMIN methodology was developed for patient-reported outcome measures, and as such, the participant number cut offs may be too stringent for direct measure evaluation where smaller participant numbers can be statistically robust [120]. Finally, risk of reporting bias was introduced by the inclusion of incidental hypothesis testing (indicative of discriminative or convergent relationships for which null findings are less frequently reported) thus the case for construct validity might have been artificially inflated.

## 5. Conclusions

Accelerometry, heartrate monitoring, direct observation, calorimetry, GPS, questionnaires and diaries have been used to assess physical activity in adults with muscular dystrophy. They were largely generalizable for adult age ranges, both genders and ambulant and non-ambulant people with a range of muscular dystrophy diagnoses. However, interpretability varied between measures and there was insufficient evidence to support their reliability, validity or responsiveness for use with adults who have muscular dystrophy. Measures identified as having most potential in this review included multi-axial accelerometry and the PASIPD questionnaire. Future evaluative studies of these, and/or other, physical activity measures for use with adults with muscular dystrophy are required. Future evaluative study design should consider direct observation as a fundamental criterion and maximizing sample size. Study design should include an awareness of activity measure limitations (in general and specific to muscular dystrophy) and the potential for improved interpretability by multi-measurement.

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## Appendix 1. Full search

1. EXP Muscular Dystrophies
2. Muscular dystrophy
3. Facioscapulohumeral
4. Limb girdle muscular dystrophy
5. Becker's muscular dystrophy
6. Myotonic dystrophy
7. Sarcoglycanopathy
8. Duchene muscular dystrophy
9. 1 or 2 or 3 or 4 or 5 or 6 or 7 or 8
10. Wheelchair
11. 9 or 10
12. Physical activity
13. EXP human activities/ or "activities of daily living"/ or EXP social participation/ or EXP exercise/ or EXP circuit-based exercise/ or EXP cool-down exercise/ or EXP muscle stretching exercises/ or EXP physical conditioning, human/ or EXP plyometric exercise/ or EXP resistance training/ or EXP running/ or EXP swimming/ or EXP walking/ or EXP warm-up exercise/ or leisure activities/ or recreation/ or dancing/ or gardening/ or EXP sports/ or EXP athletic performance/ or EXP physical endurance/ or EXP physical fitness/ or baseball/ or basketball/ or EXP bicycling/ or boxing/ or football/ or golf/ or gymnastics/ or hockey/ or martial arts/ or mountaineering/ or racquet sports/ or return to sport/ or running/ or jogging/ or skating/ or snow sports/ or soccer/ or EXP sports for persons with disabilities/ or swimming/ or "track and field"/ or volleyball/ or weight lifting/ or wrestling/ or youth sports/
14. EXP Motor Activity
15. Energy expenditure
16. Exercise
17. Exercise movement techniques/ or breathing exercises/ or dance therapy/ or tai ji/ or yoga/
18. Exercise Test/ or Warm-Up Exercise/ or Cool-Down Exercise/ or Exercise Therapy/ or Plyometric Exercise/ or Circuit-Based Exercise/ or Exercise Tolerance/ or exp Exercise/ or Exercise Movement Techniques/
19. Aerobic
20. Training
21. 12 or 13 or 14 or 15 or 16 or 17 or 18 or 19 or 20
22. Measures
23. Self report
24. Test
25. Score
26. Scale
27. Ergometer
28. Accelerometer
29. Actometer
30. Pedometer
31. Treadmill
32. Questionnaire
33. Assess
34. Index
35. Level
36. MET
37. Week
38. Frequency
39. Intensity
40. 22 or 23 or 24 or 25 or 26 or 27 or 28 or 29 or 30 or 31 or 32 or 33 or 34 or 35 or 36 or 37 or 38 or 39
41. Evaluation.mp. or EXP evaluation study/
42. EXP methodology/ or EXP validation process/ or valid\$.mp.
43. Sensitivity.
44. Specificity.
45. Reliability
46. EXP reliability/ or EXP observation/ or direct observation.mp.
47. Responsiveness
48. Validity

49. EXP outcome research/ or minimal clinically important difference.mp.
50. 41 or 42 or 43 or 44 or 45 or 46 or 47 or 48 or 49
51. 11 and 21 and 40
51. 50 and 51
52. EXP Health behaviour/ or EXP exercise/ or EXP Physical Activity/ or exercise behaviour.mp.
53. Workload.mp. or EXP Workload/
54. EXP resistance training or resistance.mp.
55. Effort.mp. or EXP exercise
56. Speed.mp. or EXP velocity/
57. Heart rate.mp. or EXP heart rate/
58. Pulse rate/ or Pulse watch.mp. or heart rate/
59. Heart monitor.mp.
60. EXP accelerometer/ or Ankle step watch.mp
61. EXP ambulatory monitoring/ or EXP Actimetry/ or EXP accelerometer/ or actigraph.mp.
62. Actometer.mp.
63. Muscle metabolism/ or EXP oxygen consumption/ or metabolism/ or oxidative capacity.mp.
64. EXP energy metabolism/ or EXP energy expenditure/ or metabolic energy expenditure.mp. or walking/
65. Movement tracking.mp.
66. GPS.mp.
67. Training log.mp.
68. Daily life activity/ or activity log.mp.
69. Training Diary.mp.
70. Physical activity/ or physical performance/ or activity diary.mp. or questionnaire/
71. EXP resistance training/ or EXP exercise intensity/ or EXP exercise test/ or EXP bicycle ergometry/ or rating of perceived exertion.mp. or EXP rating scale/
72. Borg.mp.
73. EXP rating scale/ or EXP visual analog scale/ or numerical rating scale.mp.
74. Semi structured interview/ or structure interview/ or interview.mp. or telephone interview/ or unstructured interview/ or interview/
75. Frenchay Activities Index.mp.
76. Checklist Individual Strength.mp.
77. Sickness Impact Profile.mp. or Sickness Impact Profile/
78. Activity Card Sort.mp.
79. Daily observation of activity.mp.
80. Bouchard.mp.
81. PASIPD.mp. or Physical Activity Scale for Individuals with Physical Disabilities.mp.
82. IPAQ.mp. or EXP reproducibility/ or EXP physical activity/ or questionnaire/ or EXP validation study/ or EXP walking/ or self report/
83. International Physical Activity Questionnaire.mp.
84. GLTEQ.mp. or Godin Leisure Time Exercise Questionnaire.mp.
85. Oxidative capacity.mp.
86. Physical self description questionnaire.mp.
87. 52 or 53 or 54 or 55 or 56 or 57 or 58 or 59 or 60 or 61 or 62 or 63 or 64 or 65 or 66 or 67 or 68 or 69 or 70 or 71 or 72 or 73 or 74 or 75 or 76 or 77 or 78 or 79 or 80 or 81 or 82 or 83 or 84 or 85 or 86
88. 51 and 87
89. Remove duplicates from 88.

## Appendix 2. Data extraction tool

Phase 1/2		
Author(s)		
Title		
Year		
Study Design	Pilot, feasibility, RCT, pre-post, measure evaluation, observational, cross sectional	
Study main purpose	was study primarily aimed at testing an outcome measure of PA? Y/N	
Sample population being measured	BMD, FSHD, LGMD, DM, DMD unspecified MD Rare (Myoshi, Bethlem, Laminin alpha, collagen 6, Emery-Dreifus etc)	
Sample size	Number with MD Details of participants not diagnosed with MD	

Aspect of physical activity (PA) measured	Indirect (standardised questionnaires or diaries, non-standardised diaries, PA subsection, non-standardised questionnaire or interview, single scale PA level) Direct (accelerometry, heart rate monitoring, direct observation (including supervised exercise testing like VO2 and workload), positioning sensors (like GPS), calorimetry (double labelled water, indirect calorimetry) Interpretability (scope in terms of FITT framework)	
Measure(s) of PA	Measurement tool(s) Exact wording from paper	
Measure administration	Was the measure used to assess free-living PA? Is the administrative burden discussed (time frame, training, cognitive ability required by participant, cost, expertise, equipment, copyright, other) and was this burden considered acceptable?	
Tool development	Concept to be measured stated (rationale and description, use of theoretical framework, intended population described, user involvement, item determination method appropriate)? Track back to referenced development paper	
Reliability	<u>Reliability (relative agreement)</u> (i) test re-test: ANOVA/t-test (or non-parametric equivalent) plus ICC, Kappa or Pearson's, Spearman's correlation coefficient, Bland and Altman relative agreement (ii) inter or intra-rater: % agreement, ICC, Kappa or Krippendorff's alpha or other statistic? <u>Measurement Error (absolute agreement)</u> (i) test re-test: ANOVA/t-test (or non-parametric equivalent), Kappa, Bland and Altman absolute agreement (ii) inter or intra-rater: % agreement, Kappa or Krippendorff's alpha or other statistic? <u>Internal consistency (for scale measures)</u> : Cronbach's alpha or other statistic (Kuder-Richardson formula 20, KR-20) <b>Was the test robust?</b> (interval between tests, adequate sample size, adequate results reported, reported by scale category if appropriate, appropriate statistics ICC or kappa $\geq 0.4$ , Krippendorff's alpha $\geq 0.8$ , alpha or KR-20 $>0.7$ )	
Responsiveness	What was the reported sensitivity to change or responsiveness? Cohen's d effect size, standardised response mean (SRM), Guyatt's. If scoring or cut-offs are part of data analysis, does the paper provide sufficient detail including information on response options, scoring cut offs and interpretation? <b>Was this test robust?</b> (interval between tests, change over time or change following intervention clearly reported, standardised response means, effect size, other statistic, adequate results reported)	
Validity	<u>Criterion Validity</u> Concurrent validity (correlation with another measure of PA) What was the gold standard or other PA measure referenced? Statistics reported: ICC, regression, Pearson's, spearman's, t-test, % agreement or kappa, sensitivity/specificity table for dichotomous test Predictive validity (correlation with another measure of PA at a future time point) as above <u>Construct Validity</u> Hypothesis testing: Discriminative (extreme group difference), what were the different groups? ANOVA, T-test (or non-parametric equivalent e.g. Kruskal Wallance), mean (median) difference between groups. Convergent (correlation with a related construct to PA) what was the comparison measure? ICC, % agreement, regression, Pearson's, spearman's, kappa, Bland and Altman Divergent (lack of correlation with unrelated construct to PA, sometimes referred to as discriminant - considered difficult to ascertain what might be unrelated in health research for such a broad construct as PA therefore not extracted) low correlation in above tests. Structural validity (for scale measures only): Internal validity, what was the result of the factor analysis (eigenvalue, coefficient, % total variance)? Cross cultural validity: What languages or settings are being compared (Bland and Altman, % agreement, Kappa) <u>Content Validity</u> : Were experts consulted or a formal content validity ratio/index established and what was it? Face validity assessed? Y/N <b>Was this test robust?</b> (adequate sample size, reported by scale category if appropriate, appropriate statistics ICC $\geq 0.4$ , $r \geq 0.5$ , kappa $\geq 0.4$ t-test $p > 0.05$ , $t > 1$ , adequate results reported) Was this internal validity test robust? (adequate sample size 5 per item, reported by scale category if appropriate, appropriate statistics eigenvalue $\geq 1$ , factor loading high $>0.6$ , low $<0.4$ , coefficient $\geq 0.5$ , adequate results reported)	
COSMIN generalizability	1. Median or mean age (with standard deviation or range) 2. Distribution of sex (conservatively estimated by mean $\pm 1$ SD to account for potentially skewed lower age distributions, particularly in DMD) 3. Important disease characteristics (e.g. severity, status, duration) and description of treatment 4. Setting(s) in which the study was conducted (e.g. general population, primary care or hospital/rehabilitation care) 5. Countries in which the study was conducted 6. Language in which the HR-PRO instrument was evaluated 7. Method used to select patients (e.g. convenience, consecutive, or random) 8. Percentage of missing responses (response rate)	
COSMIN Interpretability	1. Percentage of missing items 2. Description of how missing items were handled 3. Distribution of the total scores 4. Percentage of participants with lowest possible score (floor effect) 5. Percentage of participants with the highest possible score (ceiling effect) 6. Scores and change scores (i.e. means and SD) for relevant (sub) groups, e.g. for normative groups, subgroups of patients, or the general population 7. Minimal Important Change (MIC) or Minimal Important Difference (MID) Cohen's d effect size, standardised response mean (SRM), Guyatt's	
COSMIN checklist	All relevant boxes A-I	
COSMIN score	Excellent, good, fair or poor, Lowest score counts	
Comments		

**Table 3a. List of 63 measures used to assess adults with muscular dystrophy (27 indirect and 36 direct measures)**

Measure type (n)	Measure	Description of measurement (FITT parameters*)	Timeframe (mode) Environment	Metric (reported range)	Participant mobility (n)
<b>Indirect</b> standardised questionnaires and diaries (9)	IPAQ (International Physical Activity Questionnaire)	Collected activity time and frequency in minutes per day for types, including walking, transport, household, leisure and work, and intensity levels (moderate/vigorous) to yield overall estimated total energy expenditure [29,31,34-37]. (FITTO)	7 days (recall) Free-living	MET minutes per week (0-15,918)	Ambulant (462) Assisted (78) Wheelchair (125) N/R (417)
	PASIPD (Physical Activity Scale for Individuals with Physical Disabilities)	Collected activity frequency and time in hours/days per week for 13 activity types, including work, exercise and household, each allocated intensity levels multiplied to yield an overall estimated energy expenditure score [32,39,42]. (FITTO)	7 days (recall) Free-living	MET hours per day (0-39.4†)	Ambulant (20) Wheelchair (22) N/R (272)
	B-PAR (Bouchard Physical Activity Record)	Recorded activity types (categorised by intensity levels 1-9) at a frequency of every 15 minutes over 24 hours to yield an overall estimated total energy expenditure score [33]. (FITTO)	3 days (real-time) Free-living	Kcal per day (1400-4400)	N/R
	EPIC (EPIC-Norfolk Physical Activity Questionnaire)	Collected time and frequency per week of listed activity types, including household, work and recreational categories, to yield hours per week for each activity and category [40]. (FTT)	1 year (recall) Free-living	Hours per week (0-8.4† hours recreational activity)	Ambulant (11)
	PASE (Physical Activity Scale for the Elderly)	Collected frequency and time (hours/days per week) for 13 activity types, including work, exercise and household, allocated intensity levels multiplied to yield an overall score [41]. (FITTO)	7 days (recall) Free-living	Arbitrary units (43-302)	Ambulant (16)
	GLTEQ (Godin Leisure Time Exercise Questionnaire)	Collected frequency of mild, moderate and strenuous activities lasting 15mins or more in 4 questions, each intensity level allocated metabolic equivalents and multiplied to yield an overall exercise activity score [36]. (FITiO)	7 days (recall) Free-living	Arbitrary units (0-119)	Ambulant (267) Wheelchair (54)
	CFS (The Canada Fitness Survey)	Collected frequency, intensity and time for home, transport, work/school and leisure activity types, carried out yearly and seasonally, yielding an overall weighted activity score [43]. (FITTO)	1 year (recall) Free-living	Arbitrary units (2.3-21.7†)	N/R
	BPAQ (Bone-specific Physical Activity Questionnaire)	Collected types of activities carried out in each year of life, and frequency per week of activities carried out in the last year. Activities are rated according to bone-load and multiplied by time and frequency to produce the score [30,32]. (FTT)	Lifetime / 1 year (recall) Free-living	Arbitrary units (0-4)	Ambulant (29) Wheelchair (34)
	FAI (Frenchay Activities Index)	Collected frequency of lifestyle activities, timing of work activities and intensity of household maintenance, each item rated 0-3 and summed to yield an overall score [38]. (FITTO)	3-6 months (recall) Free-living	Arbitrary units (19-32)	N/R
<b>Indirect non-</b> standardised physical activity diaries (8)	Electronic Diary (Activity entered into computer software)	Recorded frequency, time and type of activity by location every 30 minutes throughout the day for triangulation with direct measure [47]. (FTT)	1 day per week (real-time) Free-living	Outdoor activity time per week (14.3 hours)	Wheelchair (1)
	Activity Diary	Recorded verification of direct measures non-wear time and non-wear activities [46,54-55] or all activities for clarification of true activity versus electrical interference or other events [40]. (FTT)	3-7 days (real-time) Free-living	Non-wear time (16-114† mins)	Ambulant (32) Wheelchair (11) Bedridden (1)
	Activity Diary + DQ (Direct Quantifier: step count)	Recorded daily walking activity by logging daily step counts in a diary [53]. (FITyO)	6 months (real-time) prescribed exercise at home	Steps per day (estimated 4500-6250)	Ambulant (16)
	Log (training log)	Collected training compliance by duration and frequency per week of training sessions, carried out on an exercise bike [29,33,49,51-52] or walking [48]. (FTT)	8-14 weeks (recall) prescribed exercise at home/gym	Sessions per week (0-4 x 15-35 mins)	Ambulant (54) Assisted (4) N/R (61)
	Log + Q (training log with self-report Quantifier: RPE)	Recorded compliance by duration, frequency and type of activity (strength exercises and walking) with activity intensity quantified by Borg CR10 rating of perceived exertion (RPE) [41]. (FITT)	8 weeks (real-time) prescribed exercise at home/gym	Sessions per week (3.5 x 20 mins); RPE (light to moderate)	Ambulant (16)
	Log + DQ (training log with Direct Quantifier: HR/rpm)	Recorded compliance by duration and frequency of bike training sessions with intensity quantified by heart rate (HR) [50] and ergometer revolutions per minute (rpm) [45] recorded in log. (FITT)	8 weeks (real-time) prescribed exercise at home/gym	Sessions per week (3-4† x 30mins); rpm (60-90)	Ambulant (6) N/R (9)



Measure type (n)	Measure	Description of measurement (FITT parameters*)	Timeframe (mode) Environment	Metric (reported range)	Participant mobility (n)
<b>Indirect</b> non-standardised physical activity diaries (8) continued	Log + Q/DQ (training log with mixed Quantifiers: RPE, HR and work)	Recorded compliance by duration and frequency of walking/ bike training sessions with intensity quantified by RPE, heartrate (HR) (Uniq Pro Trainer [56]) and workload (Monark Ergometer [44] and recorded in log. (FITT)	12-16 weeks (real-time) prescribed exercise at home/gym	Sessions per week (0-4x15-31mins†); RPE (11.4-14.1†); Watts (43-99†)	Ambulant (83)
	Telephone log + Q (training log with self-report Quantifiers: RPE and repetitions)	Collected compliance by duration and frequency of strength exercises with intensity quantified by RPE (3-point scale 1 minimal, 2 moderate, 3 marked) and exercise resistance, sets and repetitions recorded in log [57]. (FITT)	7 days (recall) prescribed exercise at home	Sessions per week (2.5 x 15-20mins); sets and reps (3 x 4-8)	Ambulant (estimated 10) Assisted (estimated 7)
<b>Indirect</b> subscales of standardised questionnaires (4)	PSDQ – activity (Physical Self-Description Questionnaire)	Assessed perception of physical activity participation by Likert rating (1 false to 6 true) of statements including examples of frequency, intensity, timing and type of activities, summed to yield an overall score [75]. (FITTO)	Current perception Free-living	N/R	Wheelchair (4)
	ACS – participation ranking (Activity Card Sort)	Ranks participation (frequency or duration) in a range of activity types (e.g. social and instrumental) and intensities (e.g. high and low physical demand) depicted on cards [58]. (FITT)	N/R Free-living	N/R	N/R
	FPACQ – transport (Flemish Physical Activity Computerized Questionnaire)	Collected duration and type/intensity of transportation (active (e.g. self-propelled wheeling) versus passive (e.g. motorised)) [72]. (ITT)	7 days (recall) Free-living	Minutes of transport per day (% active) (9-226† mins (< 5%))	Wheelchair (estimated 30)
	MAQ – activity (Modifiable Activity Questionnaire)	Collected frequency, duration and type of light and strenuous intensity exercise activities undertaken, including sports, competition and swimming [72]. (FITT)	2 weeks (recall) Free-living	Sessions per week (0->3 x 0-64 mins)	Wheelchair (estimated 42)
<b>Indirect</b> single-rating activity scales (4)	PAGS (Physical Activity Grading Scale)	Collected overall activity rating, 1-6, for summer and winter (1-hardly any activity, 5- moderate exercise for 3hrs, 6 - hard exercise several times per week e.g. skiing, running) [38,48]. (FITTO)	6-month seasons / 1 year (recall) Free-living	Arbitrary rating (1-5)	Ambulant (35) N/R (70)
	ACR (Activity Change Rating)	Collected relative activity level rating from start to finish of intervention (worsened, unchanged or improved) [51-52]. (O)	12-20 weeks (recall) Free-living	worsened – improved	N/R
	Activity Increment 1-5	Collected activity changes compared to baseline on a scale of 0 – no change to 3 – new activities undertaken [123]. (O)	14 days (recall) Free-living	N/R	Ambulant (11) Wheelchair (2)
	DOA (Daily Observation of Activity)	Recorded 4 times daily a rating of activity level from 0 (no activity) to 4 (extremely active) to yield an overall score out of 16, averaged over 14 days for each participant [65]. (O)	14 days (real-time) Free-living	Arbitrary units (4.1-8.1†)	Ambulant (65)
<b>Indirect</b> non-standardised questions and interviews (2)	Interview (study specific)	Collected frequency, time and type of exercise carried out in the year following an intervention [33], between clinic visits [124], hours of sporting activity (>/< 1000 hours) pre-onset [125] and prompted contextualisation of GPS tracker data [73]. (FTT)	1 week-1 year/ pre-onset (recall) Free-living	Sessions per week (2 x 25-30 mins); n=7 > 1000 hours pre-onset	Ambulant (16) Wheelchair (1) N/R (104)
	Survey (study specific)	Collected physical activity level (PAL) on a single Likert scale from 0-10 and types of activity participated in at different times of life during school years and beyond) [122]. (ITT)	Lifetime (recall) Free-living	PAL (5-9†)	N/R
<b>Direct</b> accelerometric monitoring (10)	Pedometer (Yamax Digiwalker, uniaxial pedometer)	Recorded ambulatory activities, encompassing frequency, intensity and timing of stepping activities, expressed overall in steps per day and prescribed at 125% baseline [53]. (O)	6 months (real-time) prescribed exercise at home	Steps per day (estimated 4500-6250)	Ambulant (16)
	Pedometer (unspecified)	Recorded ambulatory activities, encompassing frequency, intensity and timing of stepping activities, expressed overall in steps per day [41]. (O)	8 weeks (real-time) prescribed exercise at home	Steps per day (2798–8331)	Ambulant (16)
	Pedometer (triaxial, Omron Walking Style Pro)	Recorded ambulatory activities, encompassing frequency, intensity and timing of stepping activities, stride length used to calculate daily overall activity in cumulative distance [29]. (O)	4-7 days (real-time) Free-living	Distance per day (1.0-4.2 km)	Ambulant (8) Assisted (4)

Measure type (n)	Measure	Description of measurement (FITT parameters*)	Timeframe (mode) Environment	Metric (reported range)	Participant mobility (n)
<b>Direct</b> accelerometric monitoring (10) continued	Ankle actometer (unspecified)	Records ankle movements overall (encompassing frequency, intensity and timing) [37]. (O)	12 days (real-time) Free-living	N/R	N/R
	Ankle Actometer (Actilog V3.0, triaxial accelerometer)	Recorded ankle movements overall (encompassing frequency, intensity and timing) averaged as accelerations per 5-minute epoch [44,67,69]. (O)	12 days (real-time) Free-living	Mean accelerations per 5 minutes (1-74)	Ambulant (264)
	Ankle StepWatch™ (Step activity monitor (SAM) biaxial accelerometer)	Recorded ankle movements overall averaged over 7 days to yield mean steps per day, per minute within peak non-consecutive 30 minutes and highest sustained 20, 30 and 60 minutes and inactivity duration percentage total time [70-71]. (TiO)	7 days and nights (real-time) Free-living	Steps/day (716 – 5412†) Peak steps per min (21-47†); Inactive (71-90%)	Ambulant (23)
	Wrist Actigraph (Motionlogger Watch, triaxial accelerometer)	Recorded wrist movement counts overall (encompassing frequency, intensity and timing), analysed in terms of zero-crossing mode (ZCM) in counts per minute or proportional integration mode (PIM) in bits per second [46]. (O)	7 days (real-time) Free-living	ZCM counts per min (5-220†); PIM bits per second (0-90,000)	Wheelchair (11) Bedridden (1)
	Actigraph (GTX3, triaxial accelerometer)	Recorded steps overall (encompassing frequency, intensity and timing) [61]. (O)	7 days (real-time) Free-living	N/R	Ambulant (11)
	SenseWear Pro (temperature sensing biaxial accelerometer armband)	Recorded arm movements and temperatures, algorithmically calculated as overall steps, and energy expenditure expressed as average kilocalories, per day [33]. (O)	3 days (real-time) Free-living	Steps per day (2200-13400) Kcal per day (1800-3500)	N/R
	Kinesense / Gene Active (triaxial accelerometers)	Records movement counts overall (encompassing frequency, intensity and timing) [68]. (O)	2 weeks (real-time) Free-living	N/R	Ambulant (296)
<b>Direct</b> heartrate monitoring (9)	Garmin Pulsewatch (Forerunner 50)	Recorded intensity of exercise in heartrate (HR) beats per minute (bpm) and downloads provided a record of frequency and duration of activity sessions [44]. (FITi)	16 weeks (real-time) prescribed exercise at home/gym	HR (109-135† bpm); sessions per week (0-3 x 29-31†mins)	Ambulant (77)
	Polar Pulsewatch (RS400)	Recorded intensity of exercise in heartrate (HR) beats per minute (bpm) and recorded timing and frequency of warm up/ work out sessions [59]. (FITi)	6 months (real-time) prescribed exercise at home	HR (90-130 bpm); sessions per week (2 x 15-25mins)	Wheelchair (1)
	Pulsewatch (make and model not specified)	Recorded frequency, timing and intensity of heartrate (HR) training zone (linked to percentage maximal oxidative capacity (%VO2 max) (Cosmed, Quark CPET) <sup>34</sup> or percent of predicated maximal heartrate (%HRM) <sup>39</sup> ) [33,58]. (FITi)	12-16 weeks (real-time) prescribed exercise at home/gym	HR (50-70% HRM, 70% VO2max); sessions per week (1-3 x 15-90 mins)	N/R
	Polar Pulsewatch (model not specified)	Recorded intensity of heartrate (HR) training zone (corresponding to percentage of maximal oxidative capacity (%VO2 max)(CPX MedGraphics)) and recorded timing and frequency of warm up/ work out sessions [51-52]. (FITi)	32 mins - 12 weeks (real-time) prescribed exercise at home	HR (65% VO2max) sessions per week (2-5 x 35mins)	N/R
	Polar Pulsewatch (S610)	Recorded intensity of heartrate (HR) training zone (corresponding to percentage of measured or predicted maximal heartrate (HRM)) and downloads provided a record of frequency and duration of activity sessions [29,48]. (FITi)	14-16 weeks (real-time) prescribed exercise at home/gym	HR (60-87†% HRM); sessions per week (2-3 x 20-44† mins)	Ambulant (43) Assisted (4)
	Polar Pulsewatch (Accurex)	Recorded intensity of heartrate (HR) training zone (corresponding to percentage of maximal oxidative capacity (%VO2 max) (CPE MedGraphics)) and downloads provided a record of frequency and duration of activity sessions [49-50]. (FITi)	12 weeks (real-time) prescribed exercise at home	HR (65% VO2max) sessions per week (2-5 x 30mins)	Ambulant (11) N/R (9)
	Polar Pulsewatch (FT40)	Recorded intensity of heartrate (HR) training zone (corresponding to percentage of maximal oxidative capacity (%VO2 max)) and downloads provided a record of frequency and duration of activity sessions [45]. (FITi)	10 weeks (real-time) prescribed exercise at home	HR (70% VO2max) Sessions per week (3-4 x 10-30mins)	Ambulant (6)
	Polar Pulsewatch (Vantage XL)	Recorded continuous heartrate, analysed by time, frequency and overall activity above flex heartrate (HR) (established individually from rest and low speed treadmill walking test heartrate + 10 beats per minute) [53-55]. (FITiO)	3 days (real-time) Free-living	Active minutes per day (29-232†mins)	Ambulant (37)

Measure type (n)	Measure	Description of measurement (FITT parameters*)	Timeframe (mode) Environment	Metric (reported range)	Participant mobility (n)
<b>Direct</b> heartrate monitoring (9) continued	Polar heart monitor (model not specified)	Recorded continuous heartrate, analysed by time, frequency and overall activity above flex heartrate (HR) (established individually from rest and treadmill walking tests; flex HR = 87-102† bpm) and percentage maximal heartrate (HRM) [40]. (FITiO)	3 days (real-time) Free-living	Active per day (8-251mins); at 40% and 70% HRM (8-24mins and 1min)	Ambulant (11)
<b>Direct</b> observation (7)	Step counting	Observed type (walking) with frequency and intensity indicated by number of steps counted within timeframe [61]. (FITT)	7 days (real-time) Laboratory testing	N/R	Ambulant (11)
	Supervision (Supervised exercise)	Observed frequency, duration and type of exercise (tailored [58], strength [65] or ergometer cycling [51]) carried out in the gym or during home visits, with intensity of cycling rated vicariously as easy, hard or maximal [29] or resistance [65]. (FITT)	8-52 weeks (real-time) prescribed exercise at home	Cycling intensity (easy-maximal) Sessions per week (3-4† x 21-34mins)	Ambulant (8) Assisted (4) N/R (estimated 25)
	Supervision + Q (Supervised exercise with Quantifier: RPE)	Observed frequency, duration and type of class exercises (warm up, flexibility, strength, balance, aerobic and cool down exercises), with participant-rated intensity between 1 (much too easy), 3 (sufficient) and 5 (far too strenuous) [48]. (FITT)	14 weeks (real-time) prescribed exercise at gym	Exercise class intensity (3-4); sessions per week (2 x 60mins)	Ambulant (35)
	Supervision + DQ (Supervised exercise with Direct Quantifier: heartrate (HR))	Observed frequency, duration and type of exercise (treadmill walking and ergometer cycling) with intensity quantified by ECG HR monitoring (and brachial pulse palpation [74] (corresponding to % HRM [66] and %VO2 max [74]). (FITT)	12 weeks – 6 months (real-time) prescribed exercise at home/gym	Walking/cycling HR (70% VO2max / 70-85% HRM); sessions per week (4 x 15-30mins)	N/R
	Supervision + Q/DQ (Supervised exercise with mixed quantifiers: RPE, work, HR)	Observed frequency, duration and type of exercise (cycle ergometer pedalling) with intensity quantified by rating or perceived exertion (RPE), ergometer workload and heartrate (HR) (linked to percentage maximal oxidative capacity (%VO2 max) or maximal HR) [44,62-64]. (FITT)	40-42 mins – 16 weeks (real-time) prescribed exercise at home/ laboratory	RPE (11-17†); Watts (43-109†); HR (95-177† bpm) sessions per week (0-3 x 20-42mins)	Ambulant (106) Wheelchair (2) N/R (4)
	Video + DQ (Supervision via Skype with Direct Quantifier: workload)	Observed frequency, duration and type of exercise (arm cycling and strength exercises), with intensity quantified by ergometer workload in Kilocalories per minute and Theraband 50% lengthening [59]. (FITT)	8 weeks (real-time) prescribed exercise at home	Work in Kcal per minute (4); sessions per week (4 x 15-30mins)	Ambulant (65) Wheelchair (1)
	Spectating + Q (Observation of competitive sport with Quantifier: RPE)	Observed frequency, duration and type of continuous wheelchair soccer play during scrimmages and game halves with intensity measured by rating of perceived exertion (RPE) Borg 6-20 [60]. (FITT)	2 days (real-time) competitive sport pitch	Wheelchair soccer RPE (9.6-16.7†) active game time (10-15min)	Wheelchair (4)
<b>Direct</b> energy expenditure measurement (8)	Indirect calorimetry (by gas analyser (K4b2))	Recorded frequency and time during training and match play spent at different intensities quantified by oxidative capacity (VO2) in metabolic equivalents (METs) measured using a telemetric portable gas analyser [60]. (FITi)	10-15mins (real time) competitive sport pitch	VO2 in METs (0.98-2.54†)	Wheelchair (4)
	Indirect calorimetry (by gas analyser (Oxycon Mobile))	Recorded frequency and time during training and match play spent at different intensities quantified by oxidative capacity (VO2) in metabolic equivalents (METs) measured using a telemetric portable gas analyser [60]. (FITi)	10-15mins (real time) competitive sport pitch	VO2 in METs (0.98-2.54†)	Wheelchair (4)
	Indirect calorimetry (by gas analyser (Quark b2, Cosmed))	Recorded frequency and time during ergometer interval cycling spent at different intensities quantified by oxidative capacity (VO2) in millilitres of oxygen per minute per kilogram of body weight, measured using an online gas analyser [64]. (FITi)	20mins (real-time) Laboratory testing	VO2 in ml/min/kg (11-37†)	Ambulant (12) Wheelchair (2)
	Indirect calorimetry (by gas analyser (2900 Metabolic System))	Recorded frequency and time during treadmill walking at 3 speeds, for each reaching 5 minutes of steady state intensity quantified by oxidative capacity (VO2) in millilitres of oxygen per minute, measured using an online gas analyser [53-55]. (FITi)	15+mins (real-time) Laboratory testing	VO2 in ml/min (579-1109†)	Ambulant (37)
	TEE by DLW (Total Energy Expenditure by Double -Labelled Water)	Recorded overall energy in Kilocalories per day calculated by double-labelled water technique monitoring excretion of stable isotopes (2H2O and H2 <sup>18</sup> O) [127]. (O)	N/R (real-time) Free-living	Energy in Kcal per day (948-1320)	N/R

Measure type (n)	Measure	Description of measurement (FITT parameters*)	Timeframe (mode) Environment	Metric (reported range)	Participant mobility (n)
<b>Direct energy expenditure measurement (8) continued</b>	TEE by dietary intake (Total Energy Expenditure by dietary intake)	Recorded overall energy in Kilocalories per day calculated by average daily dietary intake in participants who maintained a stable weight over 6 months preceding the measurement period [126-127]. (O)	3 days (real-time) Free-living and hospital-living	Energy in Kcal per day (1,033-1600†)	Wheelchair (6) N/R (26)
	TEE by HR/VO2/Diary (Total Energy Expenditure by diary/ HR/VO2)	Recorded in Kilojoules per day by heartrate (HR) monitoring data calculated into energy expenditure by individually established treadmill oxidative capacity (VO2) equivalence and factorial diary calculations for gaps in heartrate data [53-55]. (O)	3 days (real time) Free-living	Energy in KJ per day (6340-13,820†)	Ambulant (37)
	PAL (TEE/REE) (Physical Activity Level index)	Extrapolated as an index calculated from total energy expenditure (TEE) divided by resting energy expenditure (REE) measured by indirect calorimetry [53-54,126-127]. (O)	3 days (real time) Free-living and hospital-living	PAL index (0.45-2.03†)	Ambulant (24) Wheelchair (6) N/R (26)
<b>Direct positioning systems (2)</b>	UbiTrak (GPS/Wi-Fi) (Global Positioning System and Wireless Fidelity system)	Recorded frequency, time and type (by location) of indoor and outdoor movements by wheelchair tracking system linked to Wi-Fi and GPS, augmented by an electronic activity diary, intensity quantified by distance travelled [47]. (FTT)	1 day per week (real-time) Free-living	Hours outdoors (14.3); Miles travelled outdoors (42.5)	Wheelchair (1)
	PAMS (GPS) (Participation and Activity Measurement System)	Recorded frequency, time and type (by location) of community activities (work, social, daily living and recreational) by Global Positioning System (GPS) wheelchair tracking [73]. (FTT)	14 days (real-time) Free-living	Metres wheeled (319-3795m)	Wheelchair (1)

† Range calculated by mean (or median)  $\pm$  1 standard deviation (or confidence interval) in the absence of reported range. Abbreviations: \* FITT: F – frequency; I – intensity; Ti/T – time; Ty/T – Type of physical activity; O – overall physical activity; Q – Quantifier (by additional indirect measure); D – Direct quantifier (by additional direct measure); VO2 – oxidative capacity; HR – Heartrate; HRM – maximal heartrate; ZCM – zero crossing mode; PIM – proportional integration mode; RPE – rating of perceived exertion; ECG – Electrocardiogram; MET – metabolic equivalent; TEE – total energy expenditure; DLW – double-labelled water; PAL – Physical activity level index; REE – resting energy expenditure; GPS – Global positioning system, N/R – not reported.

**Table 4a. Study characteristics of studies and evaluated measures included in phase 2**

Author	Study Description	Measure(s)	Measurement properties	COSMIN rating
Rosenberg et al., 2013	Observational descriptive cross-sectional study testing associations between depression and physical activity in 1676 people with NMD [36]	1. IPAQ (International Physical Activity Questionnaire) 2. GLTEQ (Godin Leisure Time Exercise Questionnaire)	<u>Incidental Construct validity</u> 1. and 2. Incidental convergent validity was demonstrated for both questionnaires by moderate negative associations with depression (Patient Health Questionnaire-9) (multiple linear regression, IPAQ Beta=-0.06, $r^2=0.004$ , $P=0.012$ and GLTEQ Beta=-0.13, $r^2=0.02$ , $p<0.001$ )	GOOD
Jacques et al., 2017	Observational analytic cross-sectional study of resting energy expenditure and comparison to healthy controls [30]	BPAQ (Bone-specific Physical Activity Questionnaire)	<u>Incidental Construct validity</u> Incidental discriminative validity was demonstrated by extreme group differences in BPAQ score (current) between healthy controls and adults with BMD (Independent student t-test, $p<0.05$ , mean difference in score 4.8) and between control, ambulatory and non-ambulatory adults with BMD (ANOVA, $P<0.01$ , mean difference in score 1.4)	FAIR
Heutinck et al., 2017	Observational analytic cross-sectional study of physical activity and comparison to healthy controls [72]	1. FPACQ - transportation (Subscale of Flemish Physical Activity Computerized Questionnaire) 2. MAQ - activity (Subscale of Modifiable Activity Questionnaire)	<u>Incidental Construct validity</u> 1. Incidental discriminative validity was demonstrated by extreme group differences in FPACQ transportation time and type (active/passive) between healthy controls and people with DMD (independent t-test, $p<0.001$ , mean difference 51 minutes less for healthy controls) at early and late non-ambulatory stages (independent t-test, $p<0.003$ , mean differences 48 and 84 minutes respectively less for healthy controls). In addition, extreme group differences between early ambulatory stage and late ambulatory stage for people with DMD (One-way ANOVA with post hoc tests, $p<0.022$ , mean difference 84 minutes). 2. Incidental discriminative validity was demonstrated by extreme group differences in MAQ light and strenuous activities by people with DMD at various stages of disease progression from early ambulatory to late non-ambulatory (One-way ANOVA with post hoc tests, $p<0.01$ , mean difference 29% and 34% more doing no light and strenuous exercise respectively)	FAIR



Author	Study Description	Measure(s)	Measurement properties	COSMIN rating
Janssen et al., 2016	Randomised, controlled trial secondary analysis for measure evaluation of MRI fat fraction analysis [67]	Ankle Actometer (Actilog V3.0, triaxial)	<u>Incidental Construct validity</u> Incidental convergent validity of ankle actometer (in average accelerations per 5 minutes over 12 days) with Magnetic Resonance Imaging (MRI) fat fraction of thigh musculature was demonstrated by a moderate negative correlation (Pearson's $r^2=0.27$ , $p=0.0013$ )	FAIR
Morse et al., 2016	Observational analytic cohort study of bone health in different muscular dystrophies and comparison to healthy controls [32]	1. BPAQ (Bone-specific Physical Activity Questionnaire) 2. PASIPD (Physical Activity Scale for Individuals with Physical Disabilities)	<u>Incidental Criterion validity</u> 1. and 2. Incidental concurrent validity was demonstrated between BPAQ and PASIPD by a strong association (reported in a later study <sup>31</sup> ) between the 2 questionnaires (Pearson's $r=0.71$ , $p<0.005$ ) <u>Incidental Construct validity</u> 1. Incidental discriminative validity was demonstrated by extreme group differences in BPAQ score between healthy controls and adults with muscular dystrophy (one-way between groups ANOVA with Bonferroni post hoc, $p<0.05$ , mean difference in score 21) and between DMD and all other groups (one-way between groups ANOVA with Bonferroni post hoc, $p<0.05$ , mean difference in score 24) 1. Incidental convergent validity was demonstrated by low associations of BPAQ scores with bone ultrasound scores from the radius and tibia (Pearson's $r=0.41-0.42$ , $p<0.01$ and $r=0.39$ , $p<0.05$ respectively). 2. Incidental discriminative validity was demonstrated by extreme group differences in PASIPD score between DMD and all other muscular dystrophies (LGMD, FSHD and BMD) (Kruskal Wallis test, with post-hoc Mann-Whitney U pairwise comparisons, $p<0.05$ , mean difference in score 13) 2. Incidental convergent validity was demonstrated by low and moderate associations of PASIPD scores with tibialis anterior cross-sectional area and grip strength (Pearson's $r=0.46$ , $p<0.01$ and $r=0.65$ , $p<0.05$ respectively).	FAIR
Smith et al., 2016	Observational analytic cohort study of the incidence, prevalence, age of onset and predictors of 5 chronic conditions in 1594 adults with long-term physical disability [31]	IPAQ (International Physical Activity Questionnaire)	<u>Incidental Construct validity</u> Discriminative validity was not demonstrated as there was no significant difference between IPAQ scores for participants with and without new onset of any chronic comorbid condition over the 3.5-year time period (independent student t-test, $t = -0.85$ to $1.69$ , $df=1173$ , $p>0.05$ , IPAQ mean score differences for hypertension = 188.3, coronary heart disease = 372.6, cancer = 26.2, diabetes = 206.8 and arthritis = 0.0).	FAIR
Voet et al., 2014	Randomised controlled, crossover trial of aerobic exercise training and cognitive-behavioural therapy on chronic fatigue [44]	Ankle actometer (Actilog V3.0, triaxial)	<u>Incidental Construct validity</u> Incidental discriminative validity was demonstrated by extreme group differences in average accelerations per 5 minutes between participants receiving Aerobic Exercise Training (AET), or Cognitive Behavioural Therapy (CBT), and those who received usual care (linear mixed model for repeated measurements, $p<0.05$ , mean differences AET 4.6, CBT 5.6 and at follow up AET 5.5, CBT 7.1)	FAIR
Kierkegaard et al., 2011b	Pilot randomised controlled trial of Friskis & Svettis Open doors exercise classes in 35 adults with Myotonic Dystrophy [48]	1. Training log 2. Direct observation of exercise classes 3. Polar Pulsewatch	<u>Incidental Construct validity</u> 1., 2. and 3. Incidental convergent validity was demonstrated for all 3 measures used as a composite to assess adherence, by good correlation with exercise self-efficacy scores (Spearman's $r=0.75$ , $p<0.001$ )	FAIR
Phillips et al., 2009	Observational descriptive cross-sectional study of activity patterns and barriers in 13 people with NMD and comparison to healthy controls [40]	1. Polar heartrate monitor (Polar Electro UK Ltd) 2. EPIC (Norfolk Physical Activity Questionnaire)	<u>Incidental Construct validity</u> 1. Incidental discriminative validity was demonstrated by extreme group differences in minutes active (above flex heartrate) between people with NMD and healthy controls on 3 week and weekend days (Mann Whitney U test, $p<0.05$ , median difference 154 minutes active) 2. Incidental discriminative validity was demonstrated by extreme group differences between people with NMD and healthy controls in hours per week spent in overall physical activity measured by EPIC (Mann Whitney U test, $p<0.004$ , mean difference 24.1 hours per week) and work-related activities (Mann Whitney U test, $p<0.002$ , median difference 21.5 hours/week). There was a significant difference in choice of mode of transport to travel 1 mile (Fisher's exact test, $p<0.02$ , frequency difference 10 more control participants chose walking). There was no significant difference in household activities or recreational hours per week between groups.	FAIR



Author	Study Description	Measure(s)	Measurement properties	COSMIN rating
Kalkman et al., 2007	Observational analytic case-control study of predictors of fatigue in 198 people with NMD [69]	Ankle Actometer (Actilog, V3.0, triaxial)	<u>Incidental Construct validity</u> Incidental convergent validity of average accelerations per 5 minutes from 12 days was demonstrated by low negative correlation with Sickness Impact Profile (Pearson's $r = -0.38$ , $p < 0.01$ , FSHD and $r = -0.38$ , $p < 0.01$ , Dystonia Myotonica) and Checklist Individual Strength – fatigue (Pearson's $r = -0.34$ , $p < 0.01$ , FSHD)	FAIR
Longmuir et al., 2000	Observational descriptive cross-sectional study of habitual physical activity, perceived fitness and participation in 987 disabled youths [43]	CFS (Modified Canada Fitness Survey)	<u>Reliability</u> Test-re-test reliability of the CFS ranged from 0.66 to 1.00 (reported from an earlier study [76]) <u>Incidental Construct validity</u> Incidental discriminative validity was demonstrated by significant extreme group differences in activity level measured by the CFS between participants with physical disabilities (including MD) and those with hearing impairments or chronic illness (such as congenital heart defects, Cystic Fibrosis, Arthritis, kidney disease) (ANOVA, $p = 0.001$ , mean difference 8.1 points on CFS, Chi Squared, $p = 0.001$ , difference of 24% fewer active) and between those with MD versus head injury and Spina Bifida (ANOVA, $p = 0.001$ , mean difference 10.05 points on CFS, Chi squared, $p = 0.001$ , difference of 31.5% fewer active).	FAIR
McCrory et al., 1998	Observational analytic cross-sectional study of energy expenditure and physical activity patterns in 26 ambulatory adults with slowly progressive NMD and comparison to healthy controls [55]	1. Diary (3-day activity and non-wear log book or Dictaphone logging with energy expenditure by factorial method) 2. TEE and PAL (established by indirect calorimetry (2900 Metabolic System) with heartrate equivalence 3. Polar XL vantage	<u>Incidental Construct validity</u> 1., 2. and 3. Incidental discriminative validity was demonstrated by extreme group differences in TEE (derived from factorial diary, calorimetry and heartrate monitoring) between healthy controls and adults with NMD, and between genders (2 factor multivariate ANCOVA, respectively $p = 0.001$ and $p = 0.007$ , mean differences = 2.61MJ/day and 2.44MJ/day). Similarly, for ACTEE and ACTEE as a percentage of TEE (2 factor univariate and multivariate ANCOVA, $p \leq 0.001$ and $p = 0.05$ , mean differences = 2.19MJ/day and 1.64MJ/day and $p \leq 0.001$ and $p < 0.0001$ , mean differences = 15% and 9%). 1. Incidental discriminative validity was demonstrated by extreme group differences between genders and compared to healthy controls in diary reported exercise duration, energy expenditure and intensity (light versus moderate) between healthy controls and NMD (2 factor multivariate ANCOVA, $p < 0.02$ , $p \leq 0.03$ , mean differences: 20mins, 0.5 MJ/day and 2.0 PAL and 55 mins, 1.26 MJ/day and 1.3 PAL respectively) 2. and 3. Incidental convergent validity was demonstrated by a significant relationship between TEE and active minutes and adiposity (multiple regression analysis, $R^2 = 0.63$ for a regression model including age, gender, TEE, minutes active and free fat mass, Partial $R = -0.49$ for active minutes, $p \leq 0.0001$ and $p = 0.002$ ) 3. Incidental discriminative validity was demonstrated by extreme group differences in active minutes and heartrate between genders and between healthy controls and NMD (2 factor multivariate ANCOVA, respectively $p = 0.001$ and $p = 0.002$ , mean difference = 132 min/day and 122min/day less, 11-12bpm).	FAIR
Andersen et al., 2017	Randomised, controlled, crossover trial comparing 8 weeks of high intensity interval training with usual care and comparison to healthy controls [29]	1. IPAQ (International Physical Activity Questionnaire) Accelerometer: 2. Pedometer (Omron Walking Style Pro Pedometer HJ-720IT-E2)	<u>Incidental Construct validity</u> 1. Incidental discriminative validity was demonstrated by extreme group differences in IPAQ (MET hours per week) between people with FSHD 1 and healthy controls (equal variance test and 2 tailed t-test or Mann-Whitney rank-sum test, $p < 0.05$ , mean difference in score 13) 2. Incidental discriminative validity was demonstrated by extreme group differences in Omron step distance (km per day) between people with FSHD 1 and healthy controls (equal variance test and 2 tailed t-test or Mann-Whitney rank-sum test, $p < 0.05$ , mean difference in score 2.4)	POOR
Barfield et al., 2016	Observational descriptive cross-sectional study of exercise intensity during power wheelchair soccer in 30 people with mobility impairments [60]	1. Indirect calorimetry (portable gas analyser: K4 b2c) 2. Indirect calorimetry (Oxycon Mobiled)	<u>Incidental Reliability</u> 1. and 2. Incidental inter-rater reliability was demonstrated by no significant difference in METS between gas analysers (K4 b2c versus Oxycon Mobiled) (independent group t-tests, $p > 0.05$ ) <u>Incidental Construct validity</u> 1. and 2. Incidental discriminative validity was demonstrated by extreme group difference in METS between rest and game play exertion (repeated measures t-test, $p < 0.01$ , mean difference = 0.46METS)	POOR

Author	Study Description	Measure(s)	Measurement properties	COSMIN rating
Andersen et al., 2013	Observational analytic cohort study of response to high intensity aerobic exercise and comparison to sedentary controls [64]	1. Indirect calorimetry (Online gas analyser: Quark b2; Cosmed) 2. Direct observation of training with Heartrate monitoring via exercise bike telemetry (Monark 939E) and Borg RPE	<u>Incidental Construct validity</u> 1. and 2. Incidental discriminative validity was demonstrated by extreme group differences in activity intensity during continuous (65% and 75% maximal oxidative capacity (VO2max)) or interval (85% and 95% VO2max) cycling, measured by: Indirect calorimetry (mean VO2 ml/min/kg): significant differences between healthy controls and people with LGMD and BMD (unpaired student t-test, $p < 0.05$ , mean differences = 11-16 and 14-18 respectively). There was no significant difference between FSHD and healthy controls. Heartrate (bpm), for which there were significant differences between people with LGMD and healthy controls (unpaired student t-test, $p < 0.05$ , mean differences = 29-38). There were no significant differences between healthy controls and people with BMD and FSHD.	POOR
Kierkegaard et al., 2011a	Observational descriptive cross-sectional study of functioning and disability [38]	1. FAI (Frenchay Activities Index) 2. PAGS (Physical Activity Grading Scale)	<u>Incidental Construct validity</u> 1. Incidental discriminative validity was demonstrated by extreme group differences in FAI score between participants with mild and severe impairment (Mann-Whitney U test, $p < 0.001$ , mean difference = 9) 2. Incidental discriminative validity was demonstrated by extreme group differences in PAGS rating between participants with mild and severe impairment (Mann-Whitney U test, $p = 0.001$ , mean difference N/R)	POOR
Van der Kooij et al., 2007	Randomised controlled, crossover trial investigating the effect of strength training and albuterol on pain and fatigue [65]	DOA (Daily Observation of Activity)	<u>Incidental Construct validity</u> Incidental discriminative validity was not demonstrated, there were no extreme group differences in DOA score between participants who reported pain/severe fatigue and those who did not (t-test, $p = 0.3$ and $p = 0.33$ ) Incidental convergent validity was not demonstrated there was no correlation between DOA score and pain/fatigue measured using Daily Observation of Pain/Fatigue (Pearson's $r = 0.06$ , $p = 0.66$ and $r = -0.05$ , $p = 0.73$ respectively).	POOR
Wiles et al., 2006	Observational analytic cohort study of falls and associated risk factors and comparison to healthy controls [70]	StepWatch™ (Ankle step activity monitor (SAM, biaxial, Cymatech, USA)	<u>Incidental Construct validity</u> Incidental discriminative validity was demonstrated by extreme group differences between healthy people and people with DM (mean differences: daily step count = 2876, $p < 0.001$ , peak 30-min step rate = 19.8, $p < 0.001$ , sustained 60-min step rate = 11.8, $p = 0.002$ , time inactive = 8.4%, $p = 0.01$ ) Incidental convergent validity was not demonstrated for mean daily step count over 7 days with strength (Spearman's rank correlation not significant, $r = 0.6$ , $p > 0.06$ ).	POOR
Dawes et al., 2006	A pilot randomised, controlled trial of a home-based exercise programme in 20 adults with NMD [41]	1. PASE (Physical Activity Scale for the Elderly) 2. Pedometer (unspecified)	<u>Incidental Responsiveness</u> 1 and 2. Incidental specificity of PASE and pedometer counts was demonstrated by no significant changes in either activity measure after the intervention (Wilcoxon, $p > 0.05$ , mean difference PASE = -11.64 (SD 38.31) and step count = -1485 (SD 2681)	POOR
Aitkens et al., 2005	Observational analytic cohort study of risk factors for cardiovascular disease and diabetes in 11 ambulatory adults with NMD and comparison to healthy controls [54]	1. Diary (3-day activity and non-wear log book or Dictaphone logging with energy expenditure by factorial method) 2. TEE and PAL (established by indirect calorimetry (2900 Metabolic System) with heartrate equivalence 3. Polar XL vantage	<u>Incidental Construct validity</u> 1. Incidental discriminative validity was demonstrated by extreme group differences in self-reported activity minutes of exercise between healthy controls and adults with NMD at baseline and follow up (Repeated measures ANOVA, $p = 0.05$ , mean difference = 34mins/day at baseline and 20mins/day at follow up) 2. and 3. Incidental discriminative validity was demonstrated by extreme group differences in indirect calorimetry derived PAL between healthy controls and adults with NMD at baseline and follow up (Repeated measures ANOVA, $p \leq 0.027$ , mean difference = 0.5 at baseline and 0.3 at follow up). Incidental discriminative validity was not significant extreme group differences for TEE alone ( $p > 0.05$ ). 3. Incidental discriminative validity was demonstrated by extreme group differences in heartrate derived active minutes between healthy controls and adults with NMD at baseline and follow up (Repeated measures ANOVA, $p = 0.037$ , mean difference (less for adults with NMD) = 70mins/day at baseline and 160mins/day at follow up)	POOR

Author	Study Description	Measure(s)	Measurement properties	COSMIN rating
Kilmer et al., 2005	Observational analytic cohort study testing the efficacy of a home-based activity and diet intervention in 20 people with slowly progressive NMD [53]	1. TEE and PAL (established by HR equivalence with indirect calorimetry (2900 Metabolic System) 2. HR (Polar XL vantage) 3. Pedometer (Yamax Digi-Walker)	<u>Incidental Construct validity</u> 1. and 2. Incidental discriminative validity was not demonstrated by extreme group differences in TEE and PAL pre and 6 months post intervention (1 way repeated-measures ANOVA, $p>0.05$ ). 3. Incidental discriminative validity was demonstrated by extreme group differences in pedometer daily step count pre and post intervention (1 way repeated-measures ANOVA, $p=0.001$ , mean difference 1,250 steps/day). 3. Incidental convergent validity was not demonstrated for pedometer daily step counts, there was no correlation with fitness measured (submaximal heartrate and VO2 testing) at 3 walking speeds (Pearson's correlation, $p>0.05$ )	POOR

Abbreviations: ICC – intraclass correlation coefficient, NMD – neuromuscular diseases; DMD – Duchenne's Muscular Dystrophy; FSHD – Facioscapulohumeral Dystrophy; LGMD – limb girdle muscular dystrophy; BMD – Becker's muscular dystrophy; DM – Myotonic Dystrophy (Dystrophia Myotonica); ZCM – zero crossing mode; PIM – proportional integration mode; SAM – step activity monitor; REE – resting energy expenditure; TEE – total energy expenditure; PAL – physical activity level (index=TEE/REE); ACTEE – Active energy expenditure (TEE minus REE); RPE – rating of perceived exertion; HR – heartrate; bpm – beats per minute; VO2 max – maximal oxidative capacity; METs – Metabolic equivalents; CBT – cognitive behavioural therapy; AET – aerobic exercise training; 6MWD – 6-minute walk distance; ANOVA – Analysis of Variance; ANCOVA – Analysis of covariance; N/R – not reported.



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